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ARTERIOVENOUS ANEURYSM OF THE LUNG

BY

CHARLES BAKER AND J. R. TROUNCE

From Guy's Hospital

Received October 21, 1948

Arteriovenous aneurysms in the lesser circulation produce a clear-cut clinical picture and illustrate again that we do not know how commonly conditions occur until they are first described. The number reported since Rodes' paper in 1938 indicate that it is by no means rare and it is a condition that can be successfully treated. It is striking, however, that although a congenital lesion, often with obvious evidence from early years, it has so far been diagnosed almost exclusively in adults. The recent interest in cyanotic congenital heart disease, which has followed the possibility of operative treatment, may well reveal further cases as it did in the examples here described. It is to stress the importance of earlier recognition of a treatable condition that we are reporting two cases and reviewing the present knowledge of this disease.

The First Case

D. C., aged 27, was referred to Guy's in September 1947 as a case of cyanotic congenital heart disease to see if he was suitable for a Blalock-Taussig operation. He was the youngest of five children and there was no history of a similar condition in his family. Cyanosis, which was gradually and steadily progressive, was noticed at the age of 4, and a year later a diagnosis of congenital heart disease was made. His school life was restricted more by medical advice than by ill-health or disability. At 14 he left school and worked first as an errand boy using a bicycle, but after a year he was forced to take less active employment. His working life finished at the age of 19 when he was a shop assistant—work that he could do without distress—and at this time he was still cycling five miles a day. One day he tripped up in the shop and fell, and this accident was mis-diagnosed by his employer as a fit. He was dismissed as a potential liability. His cyanosis, which was by now extreme, prevented his finding other employment, much as he desired and sought it, and he remained at home until September 1947, when hope of relief for his supposed congenital heart condition brought him to hospital. In this eight years there was gradual deterioration of his condition. A year before he was seen he had tried to cycle again and found he could barely manage a mile, and by the time he was admitted he could only walk 100 to 400 yards on the level. Œdema of the ankles had been almost continuous for the last four years and there was cough without sputum. In 1943 he was admitted to hospital with nose bleeding and this had continued at intervals particularly in hot weather. For the last two years he had attacks about once a month, particularly after bending down, when he felt weak " with a film over the left eye and sizzling in the ears "; there was no giddiness or loss of consciousness and the attack lasted ten minutes. Careful questioning could not elicit any story of hæmoptysis. He did not squat on his haunches when breathless or distressed.

On examination he was a spare well-built man, 5 ft. 7 in. in height and 112 lb. in weight. He was grossly cyanosed with a blotchy, pitted face as seen in seborrhæic subjects; there was very marked clubbing of fingers and toes (Fig. 1 and 2). He was breathless on slight exertion but there was no orthopnœa. His neck veins were not distended, the liver was just palpable but not tender; the lung bases did not sound wet, but there was slight pitting œdema of the ankles. The pulse was regular. The heart showed no clinical enlargement, which is not against a diagnosis of Fallot's tetralogy but with this in mind one was surprised to find an absence of any murmur; pulsation in the second left intercostal space was the only finding of note. The blood pressure was 105/80 in the arms and 170/110 in the legs. Screening showed a heart slightly enlarged (13/25 cm.) with the pulmonary conus prominent and an enlarged right ventricle. In the right lower lung field was a circumscribed shadow with a well-defined margin, calcified in its lower and

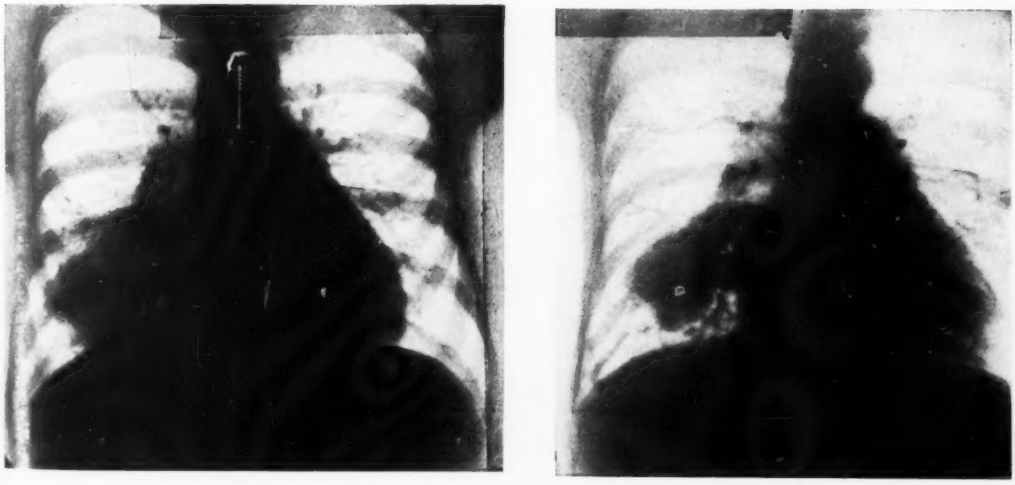
FIG. 1.—Case 1, aged 27, showing deep cyanosis and cutaneous angiomata.

lateral part. This was connected to the right hilum by a well-marked vascular shadow and there was pulsation from hilum to the tumour, which itself did not appear to pulsate; the vessels of the left hilum were not increased (Fig. 3A). The electrocardiogram (Fig. 7) showed marked right axis deviation with broad and prominent P waves in leads I and II and an inverted T in lead III.

After the shadow in the lung had been seen, the two important points needed to clinch the diagnosis of arteriovenous aneurysm were looked for, and found. On listening at the right base posteriorly there was a well-marked localized systolic murmur, but no diastolic element. Angiomata were found on the inner side of the lips and on the buttocks (Fig. 4) and it was clear the seborrhœic disfigurement of the face had masked further small angiomata though the smaller ones were still difficult to see owing to the intense cyanosis (Fig. 1). A diagnosis of arteriovenous aneurysm of the lung was made, and he was admitted in February 1948 for further investigation with a view to operative treatment.

The blood picture showed marked polycythæmia with 7·5 million red cells, a hæmoglobin of 140 per cent, and a hæmatocrit of 90; the white cells were 8000 per cu. mm. with a normal differential count. The circulation times were done by Dr. Allanby, the arm to tongue time with decholin being 14 sec. and arm to lung time with paraldehyde being 8·5 sec., both lying within normal limits. Probably the viscosity of the blood due to his polycythæmia nullified any tendency to a quickening of the circulation rate due to the shunt. The vital capacity was 2·5 litres. Unipolar limb and chest leads showed a vertical heart but no evidence of right ventricular hypertrophy. The blood volume was calculated by Dr. Reeve, the volume of red blood cells being measured by injecting a known amount of radioactive red cells. He points out that owing to the abnormal mixing of the blood the

FIG. 2.—Case 1, showing clubbing of the fingers.



A B

FIG. 3.—Case 1. (A) Straight X-ray with partly calcified aneurysm attached to right hilum by vascular shadow. (B) Angiocardiogram at 2 sec. showing two additional vascular tumours, one in the right lower zone near the mediastinum and a small one in the left lower zone behind the heart shadow.

FIG. 4.—Case 1. Buttocks showing angiomata.

figures are approximate, but a red blood cell volume of 6960 ml. and a total blood volume of 8580 ml. show an increase above normal.

The angiocardiogram taken by Dr. Hills showed well the filling of the opacity previously seen and in addition two further vascular tumours, one in the right lower zone nearer the mediastinum and a small one in the left lower zone behind the heart shadow (Fig. 3B). X-ray films of hands and feet showed no pulmonary osteopathy.

Cardiac catheterization was done by Drs. H. E. Holling and G. Zak and both branches of the pulmonary artery were entered without difficulty, and the findings are summarized in Table I.

The finding of a normal pressure in the right auricle is interesting in view of the suggestion of cardiac failure furnished by the slight œdema of his ankles. The lower pressure in the right pulmonary artery may be a consequence of the lower peripheral resistance offered by the hæmangioma on that side. The general opinion that the peripheral resistance of lung vessels is negligible is against this view, but the blood was extremely viscous and the difference in peripheral resistance of the two lungs was probably great.

Estimations of the cardiac output and of the quantities of blood passing through the pulmonary hæmangiomata were made.

Cardiac Output = Oxygen Consumption *divided by* the Oxygen Content Pulmonary Vein (Arterial Blood) *minus* Oxygen Content Pulmonary Artery (ml./litre.)

$$= \frac{230}{231 - 212} = 12 \text{ litres.}$$

Assuming that the blood passing through the pulmonary capillaries becomes 95 per cent saturated with oxygen, the pulmonary capillary circulation was estimated as 2·6 litres.

TABLE I

RESULTS OF CARDIAC CATHETERIZATION

	Inferior vena cava	Superior vena cava	Right auricle	Right ventricle	Pulmonary artery		Brachial artery
					Right	Left	
Oxygen content (Vol. per 100 ml.)	23·6	21·1	21	20·6	21·2	21·3	23·1
Percentage saturation	74	66·6	66·4	65·2	67·1	67·3	73
Pressure, mm. Hg.	8	—	8	17	12	15	105/80

The pulmonary shunt equals the cardiac output less the pulmonary capillary circulation, i.e. 9·4 (12—2·6) litres a minute, which is 80 per cent of the cardiac output.

This estimation of the proportion of blood passing through a shunt in the circulation is surprising, but is not impossible considering the depth of the cyanosis. It compares reasonably with estimated shunts in badly cyanosed congenital hearts and with arteriovenous fistulæ of the systemic circulation. The assumed figure of 95 per cent saturation of the blood after passage through the lung capillaries seems reasonable since the cyanosis of the patient did not lessen when he breathed oxygen. Even if a figure of 85 per cent saturation is assumed the shunt would still prove to be 70 per cent of the cardiac output.

Mr. R. C. Brock operated on April 5, 1948, with an anæsthetic of intratracheal cyclopropane and curare by Dr. Hutton. A right posterolateral thoracotomy found the upper and middle lobes free but the lower lobe was bound down to the chest wall and diaphragm by many collateral vessels. Some calcification could be felt on the surface and within the substance of the lower lobe, which also showed expansile pulsation and a thrill and was supplied by a very large artery, approximately 2 cm. in diameter. During the course of the operation the right middle lobe bronchus was wounded and it was therefore necessary to remove both the right lower and middle lobes. This was very difficult, partially because of the multiple vascular adhesions to the chest wall and diaphragm and partially due to the incomplete interlobar fissures, and as a result there was considerable blood loss. Before the removal of the tumour the patient's condition was poor, his systolic blood pressure was between 70 and 80 and was not improved either by transfusion or by bronchoscopic clearance of his normal lung. However, after right and middle lobes had been successfully removed he partially recovered, his blood pressure rising to 85/50 and his cyanosis disappearing. The operation lasted five hours, and during this period he received 3 pints of plasma and 1·5 pints of blood.

On returning to the ward his condition failed to improve, although his colour remained good and there was now no evidence of cyanosis; his systolic blood pressure did not rise above 90, his respirations were shallow, bubbling and stertorous, and he was deeply unconscious. X-ray of his chest showed complete collapse of his right upper lobe and on bronchoscopy a few ml. of mucoid material were aspirated. In spite of a transient improvement with this measure he lapsed into deeper coma and died some thirty hours after operation.

Necropsy was done by Dr. F. Camps, on April 7, 1948, twenty-three hours after death. There was about 250 ml. of free fluid blood in the right pleural cavity and a slight extravasation into the posterior mediastinium. The remaining upper right lobe contained no air but some lymph deposits. The left lung was moderately well ærated, deeply congested, with peripheral collapse of the lower lobe where there was also a small hæmangioma. Subpleural and pericardial hæmorrhages were seen. The heart was normal with some right sided dilatation. There was gross distension of the veins of the liver but no angiomata. The kidneys were also congested with dilated veins.

The specimen removed at operation is shown in Fig. 5 and is reported on by Dr. Allanby. "The specimen consists of the right middle and lower lobes. The pleura is wrinkled due to partial collapse of the lobes, but bears no sign of inflammation. The lateral pleural surface of the lower lobe shows patches of calcification, more easily felt than seen. Some fibrous adhesions are seen upon the diaphragmatic surface. The pulmonary vein leaving the lower lobe is grossly dilated and appears to communicate with a large thin-walled loculated system of spaces. The cut surface of the lower lobe shows a multilocular hæmangioma in its substance, measuring 3·5 cm. by 3·0 cm. adjacent to the calcified pleura, and 4 cm. from the main lower lobe bronchus. A branch of the pulmonary artery communicates directly with this cavernous space without capillary intervention. The surrounding lung and the middle lobe are normal. Sections showed the typical microscopic appearances of a cavernous hæmangioma."

The Second Case

J. A., aged 13, was admitted to Guy's Hospital in September 1948 with acute osteomyelitis in the middle of the shaft of the left femur; this was associated with a positive blood culture and quickly responded to treatment with penicillin. Cyanosis was noticed at 5 years and had progressed to a degree when it was obvious at rest, with marked clubbing of the fingers and toes. He had early been diagnosed as congenital heart disease and attended a special school, to which he was driven by ambulance; once there, he played football without any distress. Both parents and their three other children were normal. A provisional diagnosis of arteriovenous aneurysm of the lung as the cause of his cyanosis was made on admission by the registrar, Dr. W. D. Brinton, on the X-ray appearance of the lung (Fig. 6A) and was confirmed by the finding of a continuous murmur in the left upper chest as soon

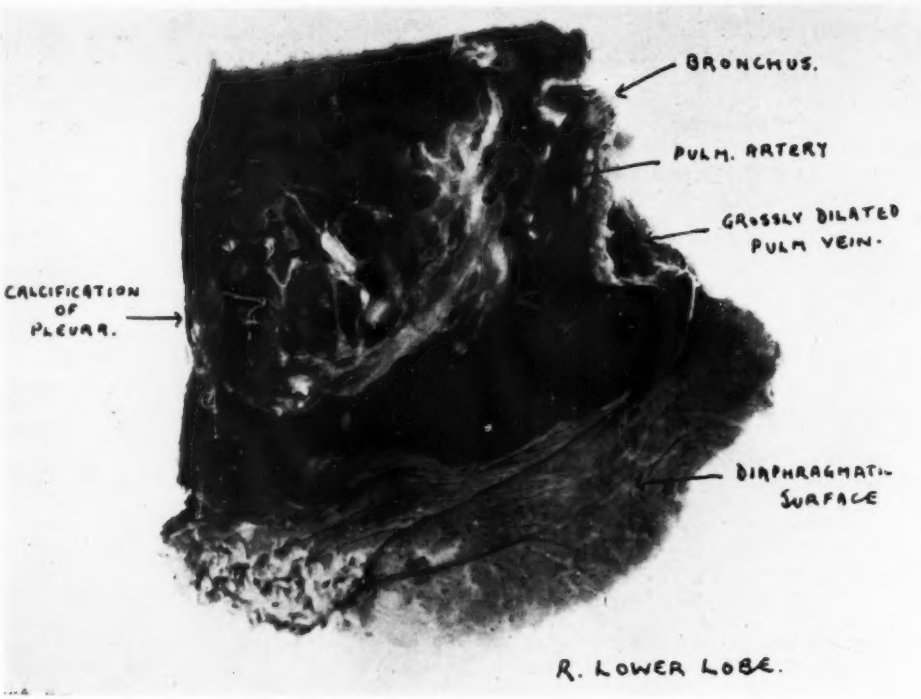


FIG. 5.—Case 1. Cut surface of lower lobe removed at operation.

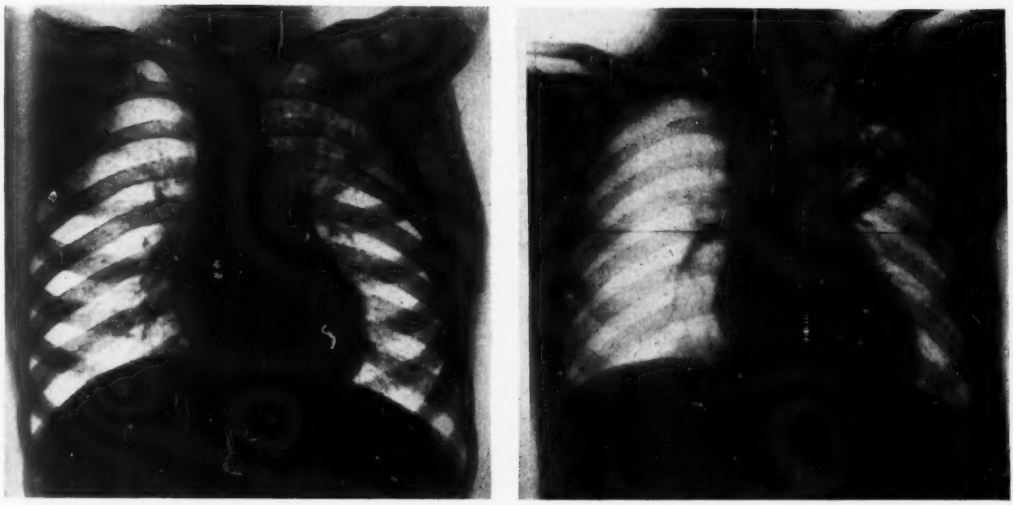
as the pulse slowed, and subsequently by angiocardiogram (Fig. 6B).

He was a boy of average height for his age, but spare. Cyanosis was obvious at rest, with some suffusion of the conjunctivæ, and was increased by exercise, though he was not unduly breathless. Clubbing was marked. He did not squat when tired. The heart was not enlarged clinically and on X-ray showed a prominent pulmonary conus only and a cardio-thoracic ratio of 10/23 cm. There was a short systolic murmur in the first and second left spaces 2 to 3 in. from the midline and an increased pulmonary second sound. Apart from this, heard over a wide area but best appreciated 10 cm. from the midline in the second space, was a more distant but clear continuous murmur, which could just be heard in the back above the left scapula; there was no thrill. The blood pressure was 115/75. Careful search found no systemic angiomata. The hæmoglobin was 126, with 7·2 million red cells and a colour index of 0·88. The electrocardiogram (Fig. 8) showed no right axis deviation. The appearance on straight X-ray was of a mottled opacity in the apical and subapical segments of the upper left lobe composed of rather worm-like streaks tending to radiate upwards and outwards from the hilum. That these were vascular shadows was clearly shown on angiocardiography where most of the dye was shunted through the opacity, the arterial elements being filled in two seconds and the larger venous channels in the third second. At the moment of reporting this case, for which we are indebted to Dr. Hampson, Mr. Brock has agreed to operate, but this has not yet been done.*

DISCUSSION

It will be seen that these two cases present a clear-cut clinical picture, but it is only of recent years that

* Mr. Brock successfully removed the left upper lobe containing the arteriovenous aneurysm on February 10, 1949; there was a large collateral circulation from the bronchial arteries. Following operation, cyanosis has disappeared and his physical capacity has increased.



A B

FIG. 6.—Case 2, aged 13. (A) Straight X-ray showing streaky radiating shadows in left upper lobe. (B) Angiocardiogram at 2 sec. from injection of dye, showing the vascular nature of the shadow in the left upper lobe.

it has been recognized. In 1936 Bowers reported a fatal case in a child of two days old, and in 1938 Rodes described the clinical picture in an adult. The first reported case treated surgically was in 1942 by Hepburn and Dauphinée, where the aneurysm was removed and the condition cured. We have found 29 cases so far reported and although they are not all described in full detail a reasonable survey can be made, to which we are adding these 2 further cases.

With the exception of the case of Bowers (1936), one mentioned by Sweet in the discussion following Maier's (1948) paper, and our second case, the diagnosis has been made in adults, the average age of 26 cases being 29, with extremes of 16 and 51. Nevertheless it appears that the condition has in most cases been present in childhood, for in 15 cases where the onset of cyanosis is given, it is since birth or early childhood in 8, between the ages of 6 and 14 in 4; and in only 3 is it stated to have begun in adult life. Both sexes are affected but in the reported cases there is a predominance of males. A family history has been suggested in five instances, of which two seem certain, and Goldman's two cases of arteriovenous aneurysm in the lung (1943 and 1947) were brothers.

Cyanosis is the most important feature and its absence was noted in only two cases, that of Duvoir and Picot (1939) who had many visceral angiomata and died of pneumonia aged 16, and Whitaker's (1947) second case, aged 33, where there was no disability; Maier (1948) makes no mention of this point in his case. It is only in these three that absence of "clubbing" is specifically mentioned, and with a few exceptions where no note is made it is, as might be expected, a feature that goes hand in hand with the degree of cyanosis. Pulmonary osteoarthropathy is reported once by Adams (1944). Polycythæmia in the cyanosed cases is a constant finding with an average of 7·3 million in red cells, the highest figure being 11·4 million in Goldman's (1943) case; the colour index tends to be low, a common finding in polycythæmia. It is likely, however, that although cyanosis is almost a constant finding in reported cases, both the increasing recognition of this condition and the increase in routine X-rays of the chest will reveal earlier cases before cyanosis develops.

It is not surprising that the combination of cyanosis, clubbing, and polycythæmia usually occurring from an early age, often suggests a diagnosis of congenital heart disease, noteworthy exceptions to this misdiagnosis being Smith and Horton's (1939) and Goldman's (1947) cases, both in adults, who had been under treatment for polycythæmia rubra vera. Nothing, however, is found amiss with the heart either on clinical or radiological examination, the sole abnormalities recorded being

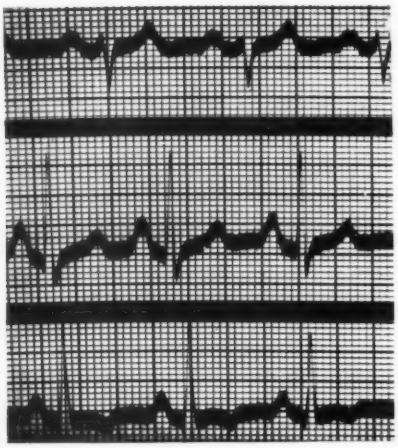


FIG. 7.—Case 1. Standard leads showing marked right axis deviation with large P waves and inverted T III.

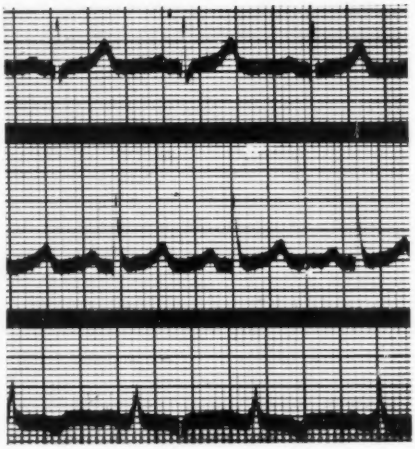


FIG. 8.—Case 2. Standard leads showing no right axis deviation.

by Sisson *et al.* (1945) with a systolic murmur and enlargement, by Alexander (1945) with enlargement to the left without a murmur, and by Lindgren (1946) whose second case had mitral stenosis. Nor, with the possible exception of Sisson's (1945) case, is there any history of these cases developing heart failure, and though congestive failure was suspected in our first case the cardiac catheterization showed that this was not present. That arteriovenous shunts in the lesser circulation produce so little effect on the heart is in marked contrast to the well recognized strain exerted when similar shunts occur in the systemic circulation. It would, therefore, seem that the combination of cyanosis, clubbing, and polycythæmia, with normal findings on examination of the heart, should suggest the possibility and search for arteriovenous aneurysm of the lung.

The main symptom of the condition is dyspnœa which has not preceded the cyanosis in any patient. The degree of disability in many has been slight, particularly considering the degree of cyanosis, and in only 7 of the 29 reported cases would it appear to have been marked. In this respect our first patient appears to have been an unusually severe and late example of the condition. Attacks of dizziness and faintness, not amounting to loss of consciousness, such as were experienced in our first case, have been noted in others. They are presumably due to the increased blood viscosity in polycythæmia and are similar to the attacks so common in morbus cœruleus, though cerebral thrombosis has not occurred. Alexander (1945), however, reports a case of a patient with marked polycythæmia who died of coronary thrombosis. Epistaxis is not uncommon and occurred frequently in our first patient, once necessitating admission to hospital. This is not unusual in subjects of polycythæmia from any cause, but might well be due, in this particular condition, to angiomata in the nasal mucosa; unfortunately we omitted to examine this site, either in life or post-mortem. Hæmoptysis is reported, and was the cause of death in Rodes' (1938) case, and Lindgren's (1946) third case had been diagnosed and treated as pulmonary tuberculosis on account of this symptom.

In contrast to the negative findings on clinical examination of the heart, a murmur in the lungs, usually systolic but occasionally with a diastolic component, was found in 14 of 23 cases where this sign is mentioned. Similarly in 19 cases where the point is made, associated lesions in other parts were found in 12, the commonest site being on the lips. These two signs, therefore, afforded strong confirmatory evidence of the diagnosis of arteriovenous aneurysm of the lung.

The clinical diagnosis is confirmed by radiology and in no case has a straight X-ray failed to show the lesion in the lung. No doubt in many instances, as in ours, the finding of a shadow away from the heart has been the finger pointing to the answer in a puzzling clinical problem. These are variously described as rounded, with defined or with irregular edges, wormlike, or as an extension of the hilar

vascular pattern; pulsation may be seen and calcification had developed in our first case. Tomography will clarify and localize, and kymography may show pulsation not visible or established by cardioscopy, but the angiocardiogram is most helpful not only in establishing the vascular nature of a doubtful shadow but in outlining aneurysms close to the mediastinum or hilum or obscured by the heart. Though the experience of Sisson *et al.* (1945) whose case died as a result of this investigation, must be remembered, an angiocardiogram was amply rewarded in our first case by showing two additional shadows not seen on the straight X-rays or on screening. As multiple shadows have occurred in about half the cases, this investigation would appear to be necessary if surgical intervention is contemplated.

Investigations of the circulatory dynamics have not been common. The circulation time in our first case was normal, which is in accordance with the findings of Makler and Zion (1946) and of Watson (1947). The percentage of blood passing through the shunt was estimated by Maier (1948) to be 58, compared with 25 to 30 in one of Lindgren's (1946) cases and the 80 in our first case. Normal intracardiac pressures were found on catheterization in our case as in Maier's (1948) but unlike our very high figure of 10 to 20 litres a minute, the cardiac output was normal. The increased blood volume which we record was found in all the 8 other estimations made. It is due to the increased cell volume, the plasma volume being unaffected, a distinction from arteriovenous aneurysms in the systemic circulation where both cell and plasma volume are increased with a normal hæmatocrit reading.

The natural course and prognosis of a condition so recently recognized and so readily subjected to surgery is not easy to estimate. From the reported cases it is clear that there is steady, though often slow progression from cyanosis alone, the first sign of a lesion of any size, to symptoms of breathlessness, to restricted activity, and in a few cases to complete disability. It must be remembered that cyanosis alone is disfiguring in the developing child or young adult and there are associated hazards from vascular accidents due to the polycythæmia or the lesion itself. In the 29 cases we have found, 5 died: one from a hæmoptysis at 25 (Rodes); one aged two days from hæmorrhage through the pleura (Bowers); one from coronary thrombosis at 41 (Alexander); one case, not cyanosed, from pneumonia at 16 (Duvoir); and one aged 45, a severe case, as the result of angiocardiography (Sisson). While the average age of the group is 19, and many had little disability except the cyanosis —notably Smith and Horton's case of a "blue baby" who at 46 had only "some dyspnœa"—there is no case older than 51. It would therefore appear that in those with lesions of any size, where cyanosis is of necessity present, the expectation of life is diminished, though up to the age of 30 the hazards are not great. This would suggest that in these, operation, if it is a reasonable risk, should certainly be contemplated, for if successful, it would mean a cure.

Even allowing for unsuccessful cases which might not so readily be reported, the results are certainly encouraging. Five of the cases died and 19 of the remaining 24 cases had operations. There were 2 fatalities, to which we now add another. In one where the operation is not described, the disappearance of cyanosis is noted as slow, but the others—three with local excision, the remainder with lobectomy or pneumonectomy—are reported as cures although post-operative observations are minimal and follow-up necessarily short. In arteriovenous aneurysm of the lung where cyanosis is present it would seem therefore that operation is not only indicated but is a reasonable risk. We know that children stand thoracic operations better than adults so that it is important to make the diagnosis at an early age. In those where the condition is recognized without cyanosis—and the quickening appreciation of it will probably reveal cases where an obscure lung shadow is the only finding—the need for surgical intervention would appear small. Whitaker's second patient who refused operation was one of these, and it is by following these types of cases that we shall get a wider appreciation of the natural course of a condition, possibly by no means rare, which so far has been diagnosed only in its more severe degrees.

Summary

Two cases of arteriovenous aneurysm of the lung are described and discussed with 29 cases previously reported. A clear clinical picture, which has only been recognized in the last ten years, is seen; and the condition is by no means rare. Cyanosis from childhood or early adult life, with clubbing and polycythæmia are the most important features. Physical signs in the heart are uncommon but a murmur may be heard in the chest, corresponding to the invariable finding of an opacity in X-ray of the lungs. Associated vascular lesions in skin, mucous membranes, and particularly the lips are common. The cyanosis is slowly progressive and is followed by dyspnœa, restricted activity, and eventually incapacity. Hazards to life are from anoxæmia, polycythæmia, and hæmorrhage from the aneurysm, rather than from heart failure. The

commonest misdiagnosis is congenital heart disease when cyanosis develops in early life, as is common, but it may also be mistaken for polycythæmia rubra vera if cyanosis first develops in adult life, or pulmonary tuberculosis when hæmoptysis occurs with the undiagnosed X-ray opacity.

It is successfully treated by removal of the aneurysm, by lobectomy or pneumonectomy. Surgery is a reasonable risk and indicated in those with lesions large enough to cause cyanosis. The importance of early diagnosis and surgical intervention at an appropriate stage in the disease is stressed.

We are indebted to the numerous colleagues at Guy's Hospital, mentioned above, who co-operated in the investigation and treatment of these two cases.

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THE HEART RATE WITH EXERCISE IN PATIENTS WITH AURICULAR FIBRILLATION

BY

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In a previous article (Knox, 1940) an accurate method of recording the changes in heart rate during exercise was described, and results were given for normal subjects performing a brief light step-test. This technique has now been applied to patients with auricular fibrillation. The present series consists of twenty-two tracings from thirteen ambulant patients with fibrillation.

The standard exercise was five ascents of two steps each ten inches high, the rate of stepping being 96 a minute. The exercise began and ended in the sitting posture and its total duration was about twenty seconds. The heart beats were electrically recorded on a smoked drum throughout exercise and were counted in five-second periods to the nearest tenth of a beat.

The clinical condition of the patients with fibrillation was classified as good, fairly good, fair, or poor on the basis of the functional capacity classification of the American Heart Association (1926). This was called the patients' "Exercise Tolerance Group." In addition, the patients were also grouped according to whether they were receiving digitalis or not.

Full details of the method of calculating the various heart rate indices were given in the previous article and only a summary need be given here.

The initial rate is the heart rate averaged over the ten seconds immediately preceding exercise.

The maximum rate is the highest rate observed during exercise averaged over a five-second period.

The acceleration of the heart rate is calculated by subtracting the rate in the five seconds immediately preceding exercise from the maximum rate, and dividing the result by the time taken to reach the maximum rate.

The post-exercise rate is the heart rate averaged over a thirty-second period beginning five seconds after exercise ends.

Results

The heart-rate indices. The figures for the various heart-rate indices of the fibrillation cases in the different "Exercise Tolerance Groups" are shown in Table I. For comparison, the mean results for 100 normal subjects (75 men and 25 women) are also included in this table. It can be seen that there was no correlation between any of these heart rate indices and the patient's clinical condition. This was not unexpected, as the number of patients in each group is very small and in addition the completely irregular ventricular rhythm present in fibrillation produces sudden arbitrary changes in heart rate in the successive five-second intervals counted. Certain general characteristics, however, can be seen in Table I.

Apart from the single case with good tolerance the maximum rates reached during the step-test were much higher than in the normal subjects. This was also true for the percentage and actual increases over the initial rates, and for the post-exercise rates.

The curve of heart rate during exercise. Fig. 1 shows the mean curves of heart rate during exercise for the different tolerance groups in all the fibrillation patients who were on digitalis. For comparison, the mean curve of the normal subjects is also included in the figure. Two of the curves (fairly good and fair) show a distinct *fall* in heart rate during the first five seconds of the exercise, a phenomenon that is of course never seen in normal subjects. In general, compared with the normal curve, the acceleration immediately after the beginning of exercise in the digitalized patients with fibrillation is less; but later during the exercise great acceleration occurs, reaching a higher value than in the normals. Thus in Fig. 1 all the fibrillation curves show a steeper slope in some portion of their course

TABLE I

Index	Mean results in 100 normal subjects	Auricular fibrillation patients grouped by estimate of their exercise tolerance					
		Good	Fairly good	Fair		Poor	
		on digitalis (1 case)	on digitalis (2 cases)	on digitalis (2 cases)	not on digitalis (1 case)	on digitalis (4 cases)	not on digitalis (3 cases)
Initial rate (beats/min.)	87	67	116	96	136	100	116
Maximum rate (beats/min.)	131	130	185	151	235	155	182
Acceleration of heart rate (beats/min./sec.) ..	2·9	2·3	2·2	2·8	6·0	3·1	2·3
Percentage increase on initial rate	53	95	60	59	78	59	64
Actual increase in beats/min.	44	63	69	56	100	56	65
Post-exercise rate (beats/min.)	93	104	140	132	176	135	165

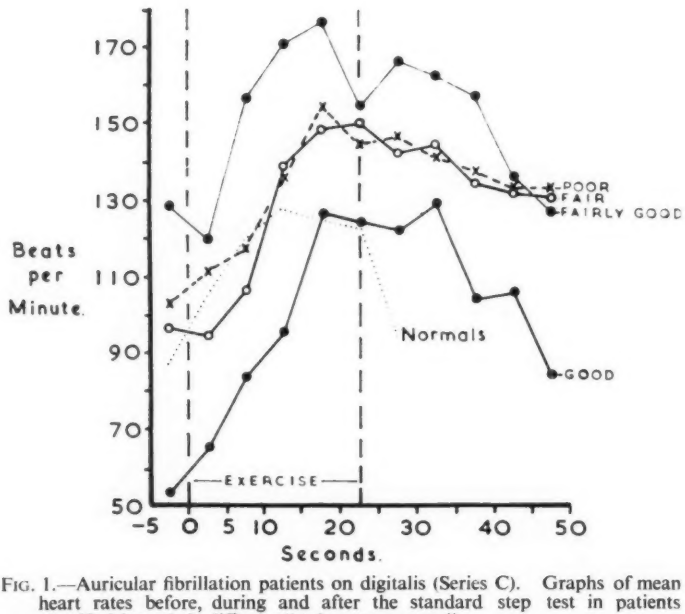

FIG. 1.—Auricular fibrillation patients on digitalis (Series C). Graphs of mean heart rates before, during and after the standard step test in patients classified into their different "tolerance groups."
The dotted line shows the mean curve for normal subjects.

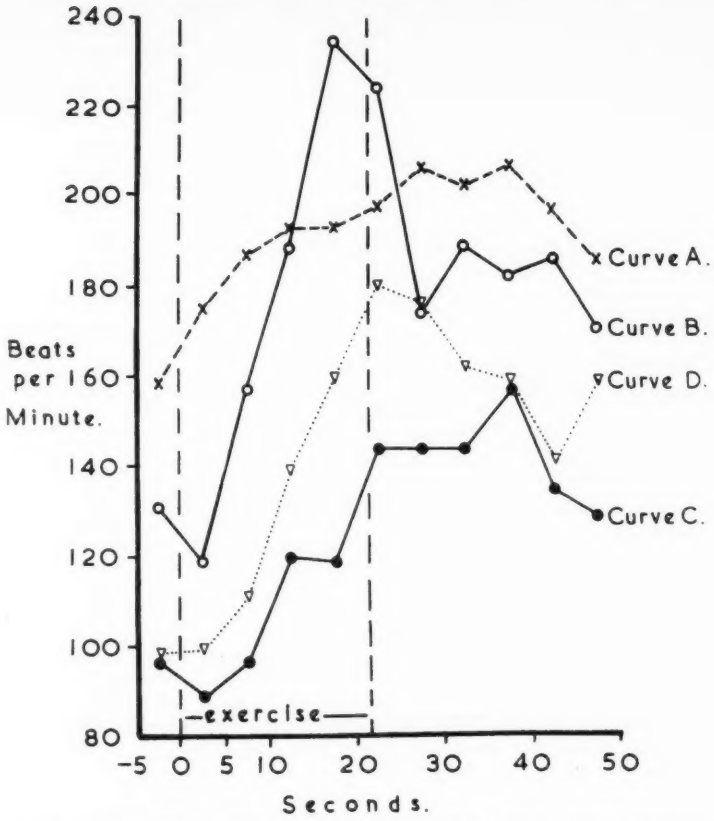

FIG. 2.—Auricular fibrillation patients not on digitalis (Series C). Graphs of heart rate before, during and after the standard step test in four patients. Curves A, B, C, and D are each from a separate patient.

than does the normal curve. This sudden delayed acceleration was very characteristic of those with fibrillation. It will also be seen that after the end of exercise the heart rate falls very slowly compared with the normal, especially in the poorer tolerance groups.

The curves of heart rate during the step test in four patients with fibrillation who were not on digitalis are given in Fig. 2. In general they show the same characteristics as the digitalized patients; these include the drop in rate at the beginning of exercise (in two cases), the delayed acceleration (in three cases) and the continuation of a high heart rate after the end of exercise. Curve B, from a patient with mitral stenosis and fair tolerance has the highest maximum rate reached by any patient, 235 beats a minute averaged over a five-second period. A portion of the actual tracing is given in Fig. 3. It is surprising that such an extreme frequency was reached during a very mild exercise lasting only twenty seconds in a patient who could get about reasonably well. In both the digitalized and non-digitalized groups the curves of heart rate for the patients with " poor " tolerance did not as a rule show such high instantaneous values of acceleration as the other tolerance groups.

Variations in a single subject. It might be expected that owing to the complete irregularity of the ventricular rate in fibrillation there would be very great variations in the shape of the heart rate curve, even in the same patient on the same day. Fig. 4 shows three curves of the response of the heart rate to the standard step test performed at 15 minute intervals by the same patient with fibrillation. The

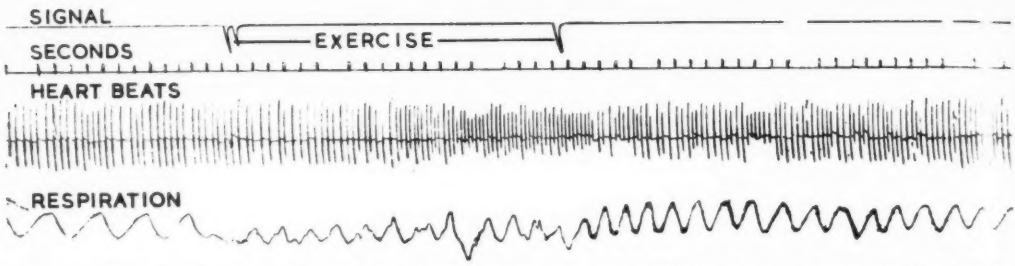

FIG. 3.—Kymograph tracing of heart beats from a patient with auricular fibrillation. Between the dips in the signal line the patient performed the standard two-step test. During the last half of exercise the extremely high heart rate of 235 beats a minute was attained.

Upper line —signal
Second line —time in seconds
Third line —heart beats
Fourth line —respiration

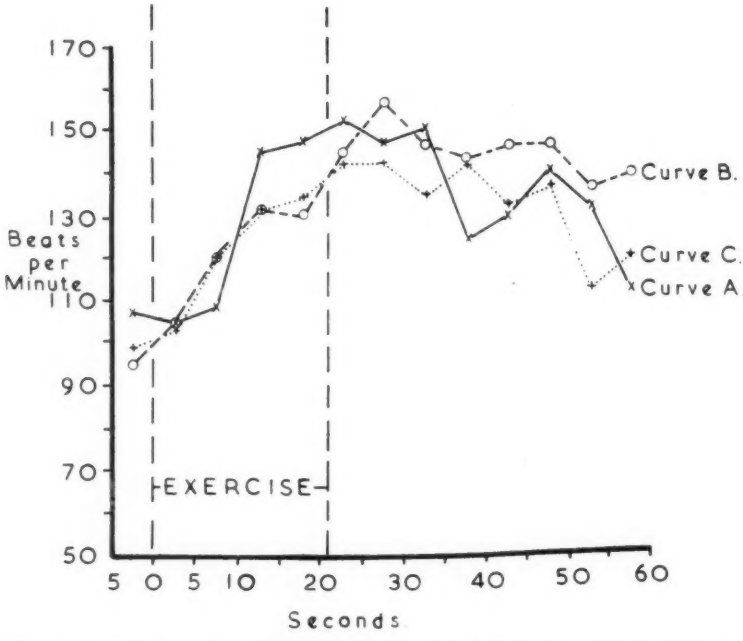

FIG. 4.—Effect of repetition of the standard step-test 3 times at 15-minute intervals on the heart rate of a patient with auricular fibrillation (Series C, Case C 49).

Curve A—Heart rate before, during, and after the standard step-test.
Curve B—The same, 15 minutes later.
Curve C—The same, 15 minutes after Curve B.

curves were taken in the order A, B, C. The variation is greater than one would expect from a normal subject, but in view of the complete arrhythmia the general shape of the curve is remarkably well maintained. It can be seen that the acceleration tends to decrease in successive tests. Similar results were obtained from two other patients with fibrillation.

There was some evidence that this constancy in the shape of the heart rate curve of a given patient may persist over considerable periods of time even when the clinical condition has improved or deteriorated. The curves for two such patients during the standard step test are shown in Fig. 5. Curve A is from a patient on digitalis whose tolerance classification was "fairly good." Curve B is from the same patient one year later when the clinical condition had improved, the dosage of digitalis remaining unchanged. Curve C shows the response in another patient with fibrillation, also on digitalis. His tolerance classification was "poor" and coupling of the beats was present. Curve D is from the same patient six months later when his general condition had become worse, though the coupling was now absent. The figure demonstrates how the characteristic shape of the curve for each patient was maintained even after an interval of many months. Thus curves A and B both show a "peak" type of curve with marked oscillations after the end of exercise, whereas curves C and D are of the "plateau" type with a relatively smooth post-exercise fall in rate.

The improvement in the condition of the first patient is accompanied by a fall in the general level of his curve, whereas the reverse is true of the second patient whose condition had become worse. While this does suggest that the exercise heart rate curves of these patients may vary with their clinical condition, it is also possible that the variations were due to different degrees of digitalization, although the actual dosage remained constant in each case.

The tracing from which curve C was taken showed the actual onset of a period of coupling. A reproduction of part of this tracing is given in Fig. 6, and the start of coupling in a burst of extrasystoles can

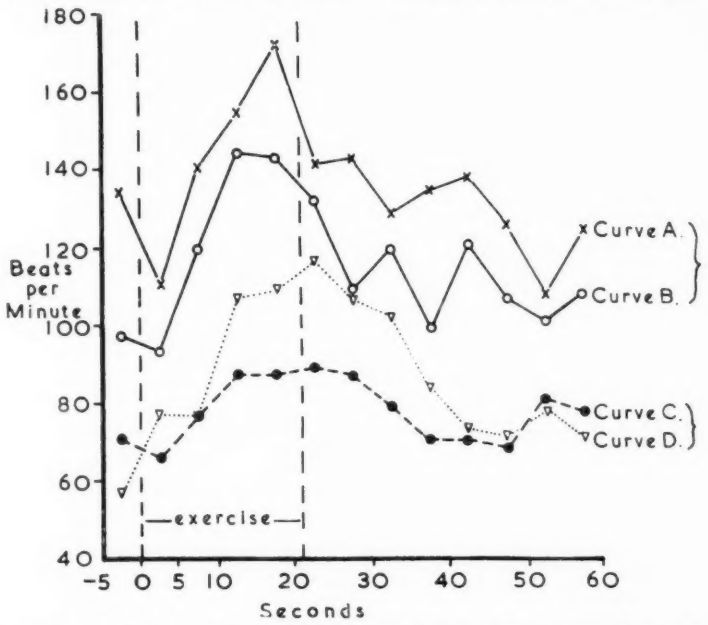

FIG. 5.—Curves of heart rate during the standard step-test in two cases of auricular fibrillation.

Curve A—Heart rate before, during, and after the step-test.

Curve B—The heart rate in the same patient one year later, when the clinical condition had improved.

Curve C—Heart rate before, during, and after the step-test in another fibrillation patient.

Curve D—The heart rate six months later in the same patient as Curve C, when the clinical condition had deteriorated.

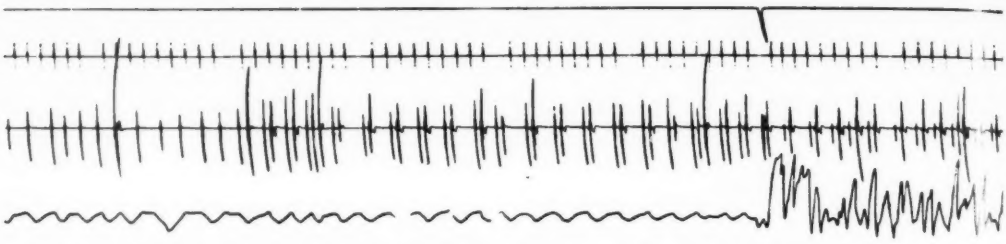

FIG. 6.—Kymograph tracing showing onset of a period of coupling in a patient with auricular fibrillation.
The coupled beats begin after a burst of extrasystoles.
The dip in the signal line indicates the beginning of the standard step-test.

Upper line —signal
Second line—time in seconds
Third line —heart beats
Fourth line —respiration

be seen to occur about thirty-five seconds before the beginning of exercise. The coupled beats persisted throughout exercise and were still present when the patient left the laboratory some fifteen minutes later. This patient may have been of the type referred to by Goodman and Gilman (1941) in whom relatively small doses of digitalis may cause coupling, as he was receiving only 1 grain tab. dig. pulv. once daily.

DISCUSSION

Blumgart (1924) was the first to attempt a quantitative estimate of the effect of exercise on the heart rate in cases of fibrillation. His exercise consisted in stepping twenty times on and off a chair, and the test was carried out by six controls and nine patients. Heart rates were recorded before and immediately after exercise, but no records were obtained during the exercise period. He concluded that fibrillating hearts respond to a given exercise by a disproportionate rise in ventricular rate and by a delayed return to normal. Both these findings are upheld by the present results as can be seen from Fig. 1 and 2.

Blumgart also found that digitalis in ordinary doses failed to prevent the exaggerated response to exercise; in fact, the actual increase in rate was slightly greater under digitalis. In the present series it was not possible to compare the results in the same patients while on and off digitalis but it can be seen from Table I that the actual increase in heart rate of the digitalized cases was much greater than in the normals so that here also digitalis has failed to protect completely against an exaggerated rise in rate. The actual increase, however, in the non-digitalized patients is on the average considerably greater than in patients of the same tolerance group who were receiving the drug. In general, if we consider the patients in the "poor tolerance" group of whom four were on digitalis while three were not, all the indices except the acceleration of the heart rate are higher in the non-digitalized group. Digitalis, therefore, while failing to abolish the exaggerated increase in rate, did appear to diminish it to a considerable extent in my cases.* Although contrary to Blumgart's findings, this is in agreement with the results of Weinstein, Plaut, and Katz (1940) who showed that digitalis when used in large therapeutic doses lessened the ventricular acceleration due to a standard exercise test in ambulant fibrillation cases.

This is of interest in connection with the work of Gold *et al.* (1939) who analysed the relative importance of the vagal and extra-vagal mechanisms by which digitalis slows the ventricle in fibrillation. They found the slowing caused by small doses of digitalis (up to about 60 per cent of the full dose) could be counteracted by large doses of atropine, so that it was largely due to vagal stimulation. When full doses of digitalis were given, atropine could no longer increase the ventricular rate and the slowing was then clearly due to extravagal actions of digitalis. The main extravagal action was said to be the increase in the refractory period of the A-V conduction system.

In 1941, Modell, Gold, and Rothendler applied these results to the exercise acceleration of the ventricle in patients with fibrillation. They concluded that in the average case the exaggerated acceleration during exercise was due chiefly, if not entirely, to decrease in vagal tone, and that blocking the vagus by atropine accelerated the ventricles to the same maximum level as extreme physical

* Dr. Maurice Campbell informs me that he (and no doubt other cardiologists) have been in the habit of teaching for years that the amount of digitalis needed for satisfactory control for a patient at rest in hospital is smaller than the amount often needed to control his heart rate when he is doing more and getting about outside the hospital. This fits in well with the experimental points.

exertion. Extra-vagal digitalization with large doses prevented the exaggerated response to exercise through direct action on the A-V conducting system, in accordance with the theory. They also pointed out that the ventricular rate at rest does not indicate whether digitalis has caused slowing by the vagal or the extra-vagal mechanism, but that there are two simple ways of detecting extra-vagal digitalization. Either 2 mg. of atropine may be given intravenously or the patient may be made to exercise. If neither of these procedures raises the ventricular rate to over 100 a minute, then enough digitalis has been given to cause slowing by the extra-vagal mechanism. Judged by the latter test, the extra-vagal mechanism can only have been prominent in one of the digitalized cases of the present series and there it was associated with marked coupling of beats. It is noteworthy that this was the only case in which no delayed acceleration of the heart occurred and it is thus probable that a sudden diminution in vagal tone was the main factor causing the delayed acceleration.

Summary

In a series of thirteen ambulant patients with fibrillation the maximum heart rates reached during a short standard step-test were much higher than in normal subjects.

No correlation was found between any of the heart-rate indices (initial rate, maximum rate, acceleration, percentage increase, actual increase, and post-exercise rate) and the clinical condition of the various patients.

In spite of the complete irregularity of the ventricular rate, the response to exercise in a given patient was reasonably constant.

In some cases a brief fall in heart rate occurred at the beginning of exercise.

An almost constant feature was a sudden delayed acceleration of the heart rate commencing about twelve seconds after exercise had begun.

Digitalis, while failing to abolish the exaggerated increase in rate, appeared to diminish it to a considerable extent.

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CALCIFIED AORTIC VALVE

CLINICAL AND RADIOLOGICAL FEATURES

BY

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The object of this paper is to suggest that a diagnosis of calcified aortic valve can justifiably be made more often than is the current practice, and that the lesion can be convincingly demonstrated and permanently recorded by a special radiological technique not hitherto described.

Rayger described the first recorded case of calcified aortic valve in 1697: he performed an autopsy, and sent one of the calcified cusps to Bonet, who described it in a publication dated 1700. A few years later Cowper (1706) described a man of forty with dyspnœa, angina, faintness and petrifaction of the aortic valve—" the valves of the Great Artery were Petrify'd insomuch that they could not approach each other." Further cases were reported by Vieussens (1715), Chevers (1842), Lloyd (1846), and Peacock (1868). Lannaec (1829) gives an excellent clinical description. The earliest extensive pathological account is that of Hasse (1846).

In more modern times, calcified aortic valve was neglected until 1931, when Christian reported a series of 22 cases, and attempted to lay down clinical criteria for the diagnosis. Interest was further aroused by the paper of Sosman and Wosika (1933) on radiological demonstration of valvular calcification, and there have been numerous American papers on all aspects of the subject up to the present time.

The English papers on the subject, however, are extremely scanty. Bennett (1930) reported a single case, and Gibbs (1935) described the pathology. Campbell and Shackle (1932) give the lesion passing mention in a general survey of aortic disease. The fullest account written in this country was that of Campbell (1937).

Frequency of the lesion. Hall and Ichioka (1940) found 31 cases of calcified aortic valve in 4000 autopsies: Dry and Willius (1939) found 23 cases in 2616 consecutive autopsies, and they further pointed out that calcified aortic valve constitutes 18 per cent of *all* healed valvular defects. The highest incidence of the lesion was that reported by Sophian (1945) who found 31 cases in 500 consecutive autopsies at the U.S. Marine Hospital. He attributed the high incidence of the lesion in his cases to age, sex, and occupation. Thus it appears that, although the available figures show a fairly wide range of variation, the lesion is undoubtedly quite common.

While discussing the incidence of the lesion, it is interesting to note how infrequently aortic stenosis occurs *without* calcification of the valve cusps. In Christian's series (1931) of 22 cases there was calcification in all but one; Dry and Willius (1939) reported 116 cases that came to necropsy, in all of which calcification was found, and 122 cases diagnosed clinically with radiologically demonstrable calcification. This shows that calcification occurs early in aortic stenosis and is almost always present by the time of death or diagnosis.

Unfortunately there are few figures available to show how frequently calcified aortic valve is diagnosed in life. Blackford and Bryan (1936) found less than 50 reported cases, that had been diagnosed in life, up to 1936. Berk and Dinnerstein (1938) reported 16 cases, of which only 5 were diagnosed in life, while Reich (1945) reported 22 cases, of which 11 were diagnosed. The later series show a much greater frequency of diagnosis than those prior to 1932: e.g. Margolis *et al.* (1931) reported 42 cases, of which only 2 were correctly diagnosed in life.

The Material for this Study

This paper describes 14 patients suffering from " pure " calcified aortic valve, in whom the lesion was suspected clinically and proved on radiological grounds. The table records the main facts concerning the patients.

Of the 14 patients, 12 were men and 2 women. Their ages ranged from 27 to 71, the average being a little over 50. Only 4 of the patients gave a

TABLE I

ANALYSIS OF FOURTEEN CASES OF CALCIFIED AORTIC VALVE

	1	2	3	4	5	6	7	8	9	10	11	12	13	14
Clinical Analysis														
Case number	1	2	3	4	5	6	7	8	9	10	11	12	13	14
Age	50	62	53	58	53	60	35	46	41	61	43	71	49	27
Sex	M	M	M	F	M	M	M	M	M	M	M	F	M	M
History of acute rheumatism	0	0	0	0	+	+	0	0	0	0	?	0	+	0
Cardiovascular symptoms	0	+	+	+	0	+	+	0	+	+	+	+	+	0
Dyspnœa	0	0	+	+	0	+	+	0	+	+	+	+	+	0
Angina	0	+	+	+	0	+	0	0	+	+	+	0	+	0
Syncope or dizziness	+	+	0	0	0	0	0	0	+	+	+	0	+	0
Œdema	0	0	0	0	0	0	+	0	0	0	0	0	0	0
Clinical cardiac enlargement	0	+	0	+	0	+	+	0	+	0	+	+	0	+
Anacrotic pulse	0	0	0	0	0	0	+	0	0	+	+	0	+	0
Blood pressure (mm. Hg.)	130	130	110	120	140	200	130	155	120	150	120	150	100	135
	100	95	80	90	85	85	100	85	80	115	90	95	80	80
Pulse pressure (mm. Hg.)	30	35	30	30	55	115	30	70	40	35	30	55	20	55
Thrill	0	0	+	0	0	+	+	+	+	0	+	+	+	+
Systolic murmur	+	+	+	+	+	+	+	+	+	+	+	+	+	+
Propagation of murmur to neck	+	+	+	+	+	+	+	+	+	+	+	+	+	+
Propagation to whole præcordium	+	0	0	0	+	0	+	+	0	+	0	+	+	+
Aortic second sound	0	0	0	0	+	0	0	0	0	0	0	0	+	+
Aortic diastolic murmur	0	0	0	0	+	0	+	0	0	+	+	0	+	+
Evidence of congestive failure	0	0	0	0	0	0	+	0	0	0	0	0	0	0
Left axis shift or L.V. strain	0	+	0	+	0	+	+	+	+	+	+	+	+	+
Wassermann reaction	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Peripheral arteriosclerosis	0	+	0	+	0	+	0	0	0	+	0	+	0	0
X-ray Analysis														
Heart size	0	+	0	+	+	+	+	+	+	+	+	+	0	+
Calcification visible on screen	+	0	+	0	0	?+	?+	0	+	+	+	+	0	?+
Calcification visible on overpenetrating X-ray	+	0	+	0	0	0	0	0	0	+	0	+	0	0
Calcification visible on tomogram	+	+	+	+	+	+	+	+	+	+	+	+	+	+

history of acute rheumatism, and in one of these it was doubtful. None of the patients gave a history of syphilis, and the Wassermann reaction was negative in all cases.

The symptoms of which the patients complained varied. Four had no symptoms referable to the cardiovascular system: of the other 10, 9 complained of dyspnœa, 8 of angina pectoris, 6 of syncope or dizziness, and 1 of œdema. One man complained of loss of memory and of the power of concentration.

The physical findings were as follows: clinical evidence of cardiac enlargement was present in 8 patients; peripheral arteriosclerosis was found in 5, and the pulse was thought to be anacrotic in 4. The blood pressure was not characteristic, but in general the systolic pressure was rather low and the diastolic rather high, unless the former were modified by associated hypertension or the latter by aortic regurgitation. The pulse pressure ranged from 30 to 115 mm. Hg., the average being 45 mm. (see Table). In 9 of the patients there was a basal systolic thrill, and in all 14 a basal systolic murmur. The basal murmur was maximal in the second right intercostal space and was propagated to the root of the neck in all cases: in 8 of the patients the murmur was audible over the whole præcordium. The aortic second sound was audible in only 3 of the cases. There was an aortic diastolic murmur in 6 cases: but the presence of such a murmur did not bear any significant relationship to the height of the pulse pressure (see Table). Electrocardiography proved of no assistance in diagnosis and showed no characteristic features: 11 patients showed evidence of left axis deviation or left ventricular strain, and of these one showed left bundle branch block, one 2 : 1 A-V block and one the typical changes of anterior coronary occlusion. The cardiographs of the 3 remaining patients showed no abnormality.

Ten of the patients have been followed up for periods up to two years. One died of congestive cardiac failure soon after admission, and 3 died suddenly within a few months of discharge from hospital: of the other 6 patients with whom contact has been maintained, one has developed severe dyspnœa and angina pectoris; the other 5 have remained unchanged.

Ætiology

The ætiology of this condition is uncertain, and this investigation adds nothing to our knowledge of the subject. There is no evidence that the lesion is due to a disturbance of calcium metabolism (Bramwell and King, 1942) to focal sepsis as suggested by Thalheimer (1922) or to syphilis (Christian, 1931; Margolis *et al.*, 1931; Grant, 1933). Libman (1913) and Perry (1936) observed that subacute bacterial endocarditis may heal by calcification, and Cohen *et al.* (1940) described one case of calcified aortic valve that showed healed aneurysms of the hepatic artery with renal and splenic infarcts. But, in general, there is no history in these patients suggestive of bacterial endocarditis.

The two views most commonly held regarding the ætiology are that it is a form of atherosclerosis (Monckeberg, 1904; Sohval and Gross, 1936), or that it is an unusual manifestation of rheumatic carditis (Cabot, 1926; Karsner and Koletsky, 1940 and 1947; Hall and Ichioka, 1940). Allbutt (1898) first pointed out the low incidence of atheroma of the aorta in cases of calcified aortic valve, and this fact has been confirmed many times since (Clawson *et al.*, 1926; Scherf, 1938); Dry and Willius (1939) make a further observation that coronary atheroma is usually in inverse proportion to the degree of aortic stenosis.

The incidence of a history of acute rheumatism or chorea in this condition is variable, the lowest incidence (4 per cent) being in the series of Friedwald and Ewing (1938), and the highest (56 per cent) in that of Cabot (1926). In the present series 4 patients (28 per cent) gave a history of acute rheumatism.

The sex distribution of the disease is remarkable, whatever the ætiology. In favour of a rheumatic origin it has been argued that chorea, which is predominantly a disease of women, is rarely followed by aortic valve disease (Contratto and Levine, 1937; Clawson *et al.*, 1938), that rheumatic carditis is a less severe disease in men than in women, and that the mild attacks are most likely to pass undiagnosed, and to be followed by slowly progressive lesions of the aortic valve (Dry and Willius, 1939).

The age incidence is more suggestive of atheroma than of rheumatic valvular disease: but cases have been recorded in young persons, and the fact that the lesion is well tolerated may lead to its escaping detection.

Histological evidence has been of little value in elucidating the problem, chiefly because authorities disagree in regard to the criteria of what constitutes evidence of old rheumatic affection.

The only conclusion warranted by the evidence available is that some patients with calcified aortic valve have, in addition, rheumatic lesions in the mitral valve; that in many cases of calcified aortic valve, without mitral valve disease, there is a history of acute rheumatism; and that those patients without mitral disease or a history of acute rheumatism, have lesions in the aortic valve that are indistinguishable from those in the other two groups.

Clinical Symptoms

Age and sex. In general the disease is one of men past middle life (Fothergill, 1879). The average age varies in different series from 52 years (Contratto and Levine, 1937) to 60 years (Friedwald and Ewing, 1938). The youngest case on record was a boy of 12 (Gautier, 1860).

The predominance of males is very striking. With the exception of Margolis *et al.* (1931), in whose series only 21 per cent were males, most series show a male incidence of over 80 per cent. This feature of the disease is difficult to explain. The suggestion that the more arduous physical life of the male, subjects the aortic cusps to greater trauma is not convincing. It seems more likely that the explanation lies in a difference in the natural history of acute rheumatism in the sexes.

Symptoms. Trousseau (1870) commented on the disparity between the mildness of symptoms and the severity of this lesion as demonstrated at autopsy. In this present series 3 of the 14 patients had no symptoms referable to the cardiovascular system.

Dyspnœa. This was the commonest symptom, occurring in 11 of the 14 cases; 2 (Cases 4 and 7) complained of dyspnœa at rest, the others only on exertion. None complained of nocturnal paroxysmal dyspnœa, which is remarkable in view of the burden thrown on the left ventricle by calcified aortic valve.

It is remarkable that the lesion may be of long duration and associated with gross cardiac enlargement without producing dyspnœa. This may possibly be explained by the slow development of the so-called "compensatory" hypertrophy of the left ventricle over a long period.

Angina pectoris. The occurrence of angina was recorded in 1706 by Cowper, whose patient "complained . . . of pain about the heart" and by Morgagni (1769). The discomfort experienced is

often atypical, conforming rather to the true etymology of the word: several of the patients (Cases 10, 11, and 13) denied retrosternal pain but complained of a choking sensation on exertion. One man (Case 3) complained of a " heavy " pain below the left nipple which was significantly related to exertion. In Cabot's series (1926), only 3 of the 9 patients with angina had typical distribution of pain. Levine (1945) also noted that the pain was often atypical, and that the history was elicited only by careful questioning.

Eight out of 14 patients in the present series (57 per cent) complained of anginal symptoms. The incidence of angina in other series varies from 7 per cent (Grant, 1933) to 60 per cent (Friedberg and Sohval, 1939). Case 4 had an attack clinically and cardiographically indistinguishable from coronary thrombosis; she did not die in hospital and no autopsy was obtained. Boas (1935) reports a patient with typical symptoms and electrocardiogram of coronary thrombosis, whose coronary arteries at autopsy were healthy; he suggests that the onset of left ventricular failure may simulate coronary thrombosis, as a result of a sudden fall in cardiac output, with reduced coronary flow.

The mechanism of angina of effort in this disease is not clear. There is rarely associated disease of the coronary arteries or their ostia (Boas, 1935; Reich, 1940; Contratto and Levine, 1937), nor can the occurrence of angina be related to aortic regurgitation. Although 3 of the 8 cases with angina had aortic diastolic murmurs, none had the peripheral manifestations of aortic regurgitation.

Friedberg and Sohval (1939) suggested that angina was due to the very high intraventricular pressure, which develops in systole with intense contraction of the ventricle, compressing the coronary vessels in a manner comparable to the blanching of the clenched fist. Levine (1945) made the ingenious suggestion that angina was due to the forcible ejection of blood into the aorta in systole, causing a suction action and leading to coronary *emptying*. But whatever the mechanism of angina may be in this condition, there is no doubt that calcified aortic valve is *the* valvular lesion above all others associated with that symptom, if luetic aortitis, which is not primarily a disease of the valve, be excluded.

Dizziness and syncope. Cowper's patient (1706) " complained of great faintness " and this appears to be one of the commonest and most disabling symptoms of calcified aortic valve. Contratto (1940) states that " dizziness and syncope help to distinguish aortic stenosis from mitral stenosis and aortic incompetence, in which these symptoms are very rare."

In the present series 5 patients complained of disturbances of consciousness varying from a transient loss of attention to complete loss of consciousness lasting for some minutes. Loss of consciousness was not accompanied by an aura, convulsions, or incontinence, and recovery was complete immediately, without disturbance of memory, speech, or motor power, and without paræsthesiæ or headache. Marvin and Sullivan (1935) and Scherf (1938), on the other hand, state that the attacks are sometimes epileptiform in character.

In some of our cases the attacks were related to exertion (Cases 1, 2, 9, and 11), a fact observed by Fothergill in 1879, but in others they occurred when the patient was at rest (Cases 10 and 13). Marvin and Sullivan (1935) related the syncopal attacks to exertion in all their 11 cases except 1, in whom the attack was post-prandial.

Mechanisms of disturbances of consciousness. It seems indisputable that the ultimate cause of loss of consciousness is cerebral anæmia, but the mechanism is obscure. Smith (1931) attributed it to insufficient left ventricular output in exertion, through a narrow aortic orifice. But in some cases syncope occurs at rest, and the occurrence of syncope is very variable, the amount of exertion which produces syncope one day having no effect on another.

There is no evidence that Stokes-Adams attacks are responsible for syncope, as the electrocardiogram between attacks shows evidence of heart block (2 : 1) in one case only (Case 11): this patient suffered from giddiness only and did not lose consciousness. Marvin and Sullivan (1935) induced an attack of syncope by exertion in one of their patients, and took continuous cardiographic tracings. At the onset of unconsciousness the heart rate was slow (50 a minute) and regular; the tracing showed nodal rhythm and marked depression of the origin of T II. A few seconds later ventricular premature beats appeared followed by simple paroxysmal tachycardia at a rate of 200 a minute. There were frequent changes in the position of the pacemaker after the subsidence of tachycardia. They concluded that loss of consciousness was due either to carotid sinus hypersensitivity, as described by Baker and Weiss (1935), or to diminished coronary flow during exertion, with a cardiac output so low as to induce unconsciousness. Carotid sinus hypersensitivity seems an improbable explanation. Contratto and Levine (1937) found that carotid sinus compression produced no effect in 19 patients with calcified aortic valve, who complained of disturbance of consciousness, and that observation has been borne out in the present series (Cases 10, 11, and 13). The most acceptable explanation so far put forward

is the occurrence of reduced coronary circulation with a marked reduction in cardiac output.

Mental changes. One patient (Case 10) complained of loss of memory and concentration sufficient to interfere with his normal activities.

Physical Signs

Blood pressure. The systolic blood pressure in the present series ranged from 100 to 200 mm.: the diastolic from 80 to 115 mm. Hg. This bears out the findings of Contratto and Levine (1937).

The pulse and pulse pressure. Lewis (1934) states: "The outstanding feature of aortic stenosis is a small pulse, rising slowly to a delayed summit: the condition should never be diagnosed without this sign." Such a view is no longer tenable. McGinn and White (1934) found a plateau pulse in only 9 of their 236 cases. In the present series the pulse was thought to be characteristically anacrotic in 4 patients only (Cases 7, 10, 11, and 13) in whom the blood pressures were 130/100, 150/115, 120/90, and 100/80 mm. Hg. respectively. In 3 others (Cases 1, 2, and 4) in whom the pulse pressure was 30 mm. or less, the pulse was thought *not* to be anacrotic. Pulse wave tracings, which might have been of value, were not obtained. In the present series the pulse pressure ranged from 30 to 115 mm., the average being approximately 40 mm. The pulse pressure presumably depends on the relative degrees of stenosis and regurgitation, and on the presence or absence of associated hypertension.

Heart size. Levine (1945) states that the heaviest hearts are those with aortic stenosis, although the largest silhouettes on X-ray examination are those with mitral stenosis. Christian (1931) found gross clinical cardiac enlargement in all his cases of calcified aortic valve, the average weight at autopsy being 680 g. Most authors agree, with the exception of Scherf (1938), who states that "cardiac enlargement does not occur until the lesion has been present for years, often several decades," and Cabot (1926) who found that the heart may be small, moderate, or large.

In the present series there was considerable enlargement of the heart in 8 of the 14 cases, slight enlargement in 3, and none in the 3 others. In 8 cases the enlargement could be found on clinical examination.

Thrill. Nine of the 14 cases had a basal systolic thrill. Reported cases show great variation in the frequency of thrills in calcified aortic valve. Thus Christian (1931) regarded the thrill as an essential criterion, whereas in McGinn and White's series of 123 cases proved at autopsy, 25 per cent had a basal systolic thrill recorded. At the other extreme Gibbs (1935) found a systolic thrill recorded in only 1 of 27 cases. It is apparent that the presence of a thrill depends on the intensity of the vibration in the heart, and on the distance of the palpating hand from the source, which may be increased by a thick chest wall or by the presence of emphysema. The low incidence of thrill in some of the series discussed from the viewpoint of pathology, such as that of Gibbs (1935), may be due to poor clinical records.

The heart sounds. Systolic murmur. Typically there is a loud systolic murmur, loudest at the base, which is conducted to the root of the neck, more on the right than on the left, and sometimes to the cardiac apex. In this series the murmur was harsh, and was propagated to the neck in all cases: in only 8 cases was it audible over the whole præcordium. In the series of McGinn and White (1934), all 123 cases had basal systolic murmurs, but the authors point out that in some of the cases which came to autopsy a harsh systolic murmur audible over the whole præcordium had, before death, been attributed to mitral valve disease. Cabot (1926) found that in 4 of his 28 cases, the systolic murmur was audible *only* at the apex.

The murmur is not characteristic (Scherf, 1938), for a similar murmur may be heard in aortic incompetence and in luetic aortitis. Nor is its propagation significant, for this depends on the intensity of the murmur (Contratto and Levine, 1937).

Diastolic murmur. Lewis (1934) requires an aortic diastolic murmur as well as an anacrotic pulse for the diagnosis of aortic stenosis, but that view is no longer held. In the present series an aortic diastolic murmur was heard in 6 out of 14 cases (42 per cent), and this inconstancy of the diastolic murmur is borne out in other series. Thus McGinn and White (1934) found diastolic murmurs in 53 per cent of cases, Contratto (1940) in 50 per cent, Cabot (1926) in 57 per cent, and Dry and Willius (1939) in 33 per cent.

Aortic second sound. On theoretical grounds, it seems unlikely that calcified aortic valve cusps could close with sufficient vigour to produce a second aortic sound. In the present series the second sound was inaudible in 11 cases and audible in 3 (Cases 5, 13, and 14), in whom it was of normal intensity. Contratto and Levine (1937) suggest that the audible second sound is really a pulmonary second sound. Scherf (1938) regards a weak or absent second aortic sound as helpful in distinguishing the murmur of calcified aortic valve from that of luetic aortitis and atheroma of the aorta, in which the sound is normal or accentuated. It may be said that whereas the absence of the aortic second

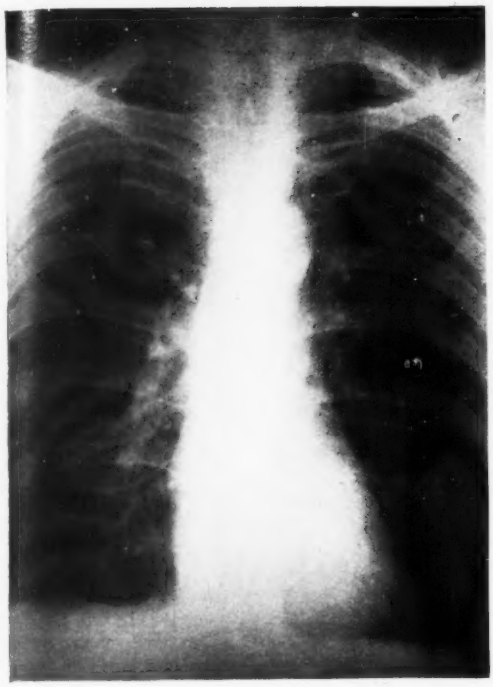

FIG. 1.—Case 1. Teleradiograph showing normal heart size and rounding of left cardiac contour, indicating left ventricular hypertrophy.

FIG. 2.—Oblique radiograph of the heart in the cadaver.
A. represents the shadow of a lead strip in the aortic valve area.
B. represents the shadow of a lead strip in the mitral valve area.

sound favours the diagnosis, its presence does not exclude it.

Evidence of congestive cardiac failure. In the present series one patient only (Case 7) presented with signs of congestive heart failure. Other authors have found the incidence much higher; for example, 26 of Cabot's 28 cases presented with œdema.

Disturbances of conduction. There have been many reports of disturbances of conduction in this condition. Cowper's case (1706) had "an intermission of one stroke in three of the pulse," and Parkes-Weber (1897) and Allbutt (1898) described cases with heart block. There have been numerous electrocardiographic reports and an extensive survey by Dry and Willius (1939). The electrocardiogram is frequently normal. The abnormalities commonly found are left axis shift, T wave inversion, A-V block which may vary in degree in the same patient (Boas, 1935), bundle branch block, smaller degrees of intraventricular block, and, very rarely, auricular fibrillation (Contratto and Levine, 1937).

Heart block has been attributed to invasion of the conducting tissue by an extension of calcification from the valve (Boas, 1935 and East, 1932). This may be so in some cases, but it would not account for intraventricular disturbances, nor for variable disturbances. Diminished coronary flow is probably the causative factor in most cases.

RADIOLOGICAL COMMENT

Postero-anterior X-rays of the chest in an "early" case of calcified aortic valve show no evidence of cardiac enlargement: the only change observed is "rounding" of the left lower border of the heart, indicating left ventricular hypertrophy (Fig. 1). The aortic shadow is normal, but a slowly expanding pulsation of the aorta may be seen on the fluorescent screen. In the later stages of the disease, left ventricular enlargement becomes obvious, with increased transverse diameter of the heart.

FIG. 3.—Oblique tomogram of the heart demonstrating the shadows of calcified mitral and aortic valve. The mitral shadow is projected below the aortic one.

FIG. 4.—Case 1. Oblique tomogram of the heart demonstrating the shadow of a calcified aortic valve.

Radiological demonstration of the calcified aortic valve is essential for accurate diagnosis. The two methods in common use are fluorescent screen examination and localized overpenetrating X-rays of the heart in oblique positions. Intracardiac calcification was demonstrated radiologically for the first time by Klason (1931), who diagnosed a calcified annulus fibrosus on the fluorescent screen: but he failed to reproduce it on an X-ray film. Saul (1932) was the first to succeed in obtaining a permanent record of a similar case. Sosman and Wosika (1933) reported 12 cases of calcified aortic valve, and 13 cases of calcified mitral valve, in which they had observed the intracardiac calcification on the fluorescent screen. In their paper a number of important technical points are stressed, among them the need for adequate dark adaptation prior to screening, the use of minimum screen aperture, and examination in the oblique positions. Many workers have reported successful application of the technique described by Sosman and Wosika (Parade and Kuhlman, 1933; Bishop and Roesler, 1934; Sparks and Evans, 1934; Sundberg, 1941; and Odquist, 1944).

In the present series demonstration of calcified valves by screen examination or overpenetrating radiographs was not possible in all cases. An attempt was therefore made to show the valves by tomography.

Tomography was carried out with the patient lying supine in the left posterior oblique position: the tube was centred 10 cm. below the suprasternal notch in the midline. Three or four horizontal "sections" were taken at 1 cm. intervals above and below an arbitrary plane, which experience has shown should be 13 to 14 cm. above the table top: some of these X-rays inevitably include both aortic and mitral valve areas.

The anatomical position of the aortic and mitral valves in X-rays taken in the left posterior oblique position was first ascertained in the cadaver, after lead strips had been placed at the level of the aortic

FIG. 5.—Case 3. Oblique tomogram of the heart: the calcified aortic valve is demonstrated.

and mitral cusps of a normal human heart (Fig. 2). Le-Wald (1916) used a similar method to demonstrate the position of the heart valves relative to the anterior thoracic wall.

Analysing the shadows cast by calcified heart valves in such tomograms, it is seen that the aortic valve is projected below and dorsal to the level of the pulmonary conus: the mitral valve lies below the aortic and roughly in the same plane (Fig. 3). Slight variations in position are common, depending on the size of the heart and associated valvular lesions. Sosman (1934) has discussed these variations extensively; the findings in the present series are similar. If the aortic valve is heavily calcified a clearly defined shadow giving the appearance of a ring is seen (Fig. 4 and 5). In other cases more vaguely outlined shadows are found in the valve area (Fig. 6 and 7).

FIG. 6.—Case 11. Oblique tomogram of the heart demonstrating calcification of the aortic valve.

FIG. 7.—Case 2. Oblique tomogram of the heart demonstrating calcification of the aortic valve. Two separate areas of calcification can be seen.

In 6 of the cases reported here, calcification of the aortic valve could be seen clearly on the fluorescent screen: in 3 there was a doubtful shadow that could not be identified with certainty: in the other 5 cases screening failed to show any evidence of calcified valves. In only 4 cases was reproduction of the calcified shadow on an overpenetrating oblique X-ray possible. Tomography showed the lesion in all 14 cases.

It appears, therefore, that tomography is the best method at present available for demonstrating and recording intracardiac calcification.

PROGNOSIS

Allbutt (1898) states: "Aortic stenosis is a long disease, for life may continue under favourable circumstances until the aperture is reduced to the size of a crow-quill or less." That point of view still holds today (Scherf, 1938).

The prognosis varies with the age of the patient, and the presence or absence of symptoms at the time of diagnosis. If discovered in a fairly young person without symptoms, the prognosis is good, the average age at death being 52 (Contratto, 1940). If, however, the patient is first seen with symptoms, the prognosis is poor, the average expectation of life in the presence of congestive cardiac failure being 9 months (Contratto, 1940); of dyspnœa, 23 months; and of syncope, 9 months (Contratto and Levine, 1937).

In the present series 4 out of 14 are known to be dead. Case 2 died within 6 months of the first attack of syncope, and within 18 months of the onset of angina at the age of 62. Case 4 died at the age of 58, having been known to have a heart murmur for 30 years, and with 5 years history of dyspnœa, and 3 years history of angina. Case 6 died at the age of 60, after one year's mild angina and dyspnœa: he had had acute rheumatism 40 years previously. Case 7 died, aged 35, after 6 months dyspnœa of effort, and œdema of the ankles for two days.

Causes of death. A certain number of cases die

from causes not connected with the cardiovascular system (as many as 40 per cent in the series of Reich, 1945).

Congestive cardiac failure. Congestive cardiac failure is the commonest cause of death: it is responsible for about 35 per cent of deaths in the series reported by Grant (1933), by Reich (1945) and by Contratto and Levine (1937). In contrast to mitral stenosis, where the patient may survive several attacks of congestive failure, a patient with failure due to calcified aortic valve rarely recovers (Contratto, 1940). One patient (Case 7) in the present series died of congestive failure after a very short illness.

Sudden death. After congestive cardiac failure, sudden, unexpected death is the commonest termination. It is of interest that the first case recorded (Bonet, 1697) describes sudden death in a Parisian tailor who was apparently in excellent health. Many other similar cases have been reported (Lloyd, 1846; Gautier, 1840; Trousseau, 1870; Wilks and Moxon, 1898; Grant, 1933; Lutembacher, 1921). The incidence of sudden death is variably estimated, the average figure being about 18 per cent, although in some series it is very much lower (Campbell and Shackle, 1933), and in others very much higher, as in the selected material of Marvin and Sullivan (1935). In the present series, of the four patients who have died in a two year follow-up, three died suddenly.

The features and cause of sudden death have been carefully studied by Marvin and Sullivan (1935). They find that death occurs in seconds rather than in minutes, resembling death from coronary thrombosis: but no disease of the coronary arteries is usually found at autopsy. Nor is the sudden death related to the degree of aortic stenosis. They suggest a relation to heart size, pointing out the relative ease with which ventricular fibrillation is induced in large hearts. The relation between syncopal attacks and sudden death is not constant, but is sufficiently frequent to suggest that both are due to a sudden reduction in coronary blood flow with low cardiac output. De Veer's suggestion (1938) that sudden death may be due to " locking of the cusps with complete occlusion of the aortic orifice " is quite unconvincing.

But, whatever the mechanism, sudden death is a frequent event in cases of calcified aortic valve, even though they are symptom-free, and therefore necessitates a guarded prognosis.

Other causes of death. A few patients have died of superadded subacute bacterial endocarditis (Contratto and Levine, 1937) and a few of coronary thrombosis (Boas, 1935).

Summary

The clinical and radiological features of 14 cases of calcified aortic valve are described, with a method of demonstrating the valves by cardiac tomography.

Twelve of the 14 cases were men, 2 were women. Their ages ranged from 27 to 71 years, the average being about 50 years. Four gave a history of acute rheumatism in the past.

The symptoms complained of were dyspnœa, angina, syncope, œdema, and loss of memory, in that order of frequency. Four patients had no cardiovascular symptoms.

The only constant physical finding was a basal systolic murmur propagated to the root of the neck; other findings were absence of the second aortic sound (11 cases), an aortic systolic thrill (9 cases), an aortic diastolic murmur (6 cases), cardiac enlargement (8 cases), and low pulse pressure (7 cases).

The electrocardiograms of 3 of the patients were normal: 11 showed left axis deviation, and of these one showed left bundle branch block, one 2 : 1 A-V block, and one anterior coronary insufficiency.

Four of the 14 patients are known to be dead in a two-year follow up: three died suddenly and one in congestive cardiac failure.

Tomography was found to be a more successful method of demonstrating calcified valves than screen examination or overpenetrating oblique X-rays.

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RIGHT BUNDLE BRANCH BLOCK AND CARDIAC INFARCTION

BY

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It is the object of this paper to investigate the modifications of the electrocardiographic pattern of cardiac infarction caused by the simultaneous presence of right bundle branch block, and to describe the cardiographic signs on which the diagnosis of infarction can be made in the presence of right branch block. The cardiogram of right branch block shows added changes characteristic of infarction; it differs in this respect from that of left branch block which often masks the evidence of infarction.

An analysis of tracings taken from patients with right bundle branch block but without infarction will be necessary for the study of cardiograms showing the combined lesion and will, therefore, precede it. All cases under review have the cardiographic pattern of the common variety of right branch block; cases of classical right branch block, characterized by a predominantly downward directed main deflection in lead I, a conspicuous R in lead II, and a tall R in lead III, with T waves in the opposite direction of the main deflection, were not included because of their rarity. A series of twenty-three patients were observed personally; the majority were seen in private practice, and their tracings were taken with a Siemens electrocardiograph; seven patients were seen at the Out-Patient Department of the National Heart Hospital, under Dr. William Evans. A search was made for reported cases to bring the total number of records up to 72.

Right Bundle Branch Block in Healthy Subjects

Right bundle branch block is not rare in healthy subjects and was found 7 times when 1445 healthy persons were examined by Wood, Jeffers, and Wolferth (1935). The present analysis is based on 20 cases (see Fig. 1 and Table, Cases 1–20). In 7 cases Q deflections were shown in lead I; they were less frequent and never large in lead II. The upstroke of the main deflection and the beginning of the downstroke were slender and steep as in normal curves. The R–T segment showed no significant depression or elevation except for a gradual rise if the following T was high and set close to the QRS complex. The T waves were upright in leads I and II in all cases. The chest lead cardiogram of right branch block, made familiar through the work of Wilson and his collaborators (1934, 1944), was examined in 14 cases, and it included leads from the right præcordial area in 11. Lead CR1 or V1 showed a bifid QRS complex, with two peaks above the isoelectric line; in some records the initial peak was of low voltage (see Fig. 1D); there were no Q waves; the R–T segment was either level with the

TABLE OF CASES

No.	Reference	Clinical Diagnosis
1.	Own case	Healthy subject.
2.	Own case	Retinal arteriosclerosis, normal heart.
3.	Own case	Healthy subject.
4.	Own case	Bronchitis.
5.	Own case	Bronchitis.
6.	Own case	Healthy subject.
7.	Own case	Healthy subject.
8.	Own case	Paget's disease.
9.	Wilson *et al.* (1934), Fig. 1	Cancer of prostate.
10.	Wilson *et al.* (1934), Fig. 2	Pleural effusion.
11.	Wilson *et al.* (1934), Fig. 3	Arthritis.
12.	von Deesten *et al.* (1934), Fig. 1	Dyspepsia.

TABLE OF CASES—*continued*

No.	Reference	Clinical Diagnosis
13.	von Deesten *et al.* (1934), Fig. 2	Extrasystoles.
14.	von Deesten *et al.* (1934), Fig. 3	Healthy subject.
15.	von Deesten *et al.* (1934), Fig. 4	Healthy subject.
16.	von Deesten *et al.* (1934), Fig. 5	Healthy subject.
17.	Wood *et al.* (1935), Fig. 1	Healthy subject.
18.	Wilson *et al.* (1944), Fig. 11	Ulcerative colitis.
19.	Wolferth *et al.* (1947), Fig. 1	Healthy subject.
20.	Goldberger (1947), Fig. 62	Healthy subject.
21.	Own case	Hypertension.
22.	Own case	Aortic stenosis.
23.	Own case	Hypertension, cerebral hæmorrhage, uræmia.
24.	Own case	Stokes-Adams syndrome and partial block.
25.	Own case	Aortic stenosis.
26.	Own case	Hypertensive heart failure.
27.	Stenström (1927), Fig. 24	Heart failure.
28.	Wood *et al.* (1935), Fig. 2	Ventricular septal defect.
29.	Evans *et al.* (1937), Fig. 4	Congenital heart disease.
30.	Evans *et al.* (1937), Fig. 5	Mitral stenosis, fibrillation, hypertension.
31.	Evans *et al.* (1937), Fig. 7	Fibrillation.
32.	Evans *et al.* (1937), Fig. 8	Hypertensive heart failure.
33.	Evans *et al.* (1937), Fig. 9B	Heart failure.
34.	Evans *et al.* (1937), Fig. 10	Hypertension.
35.	Evans *et al.* (1937), Fig. 12	Aortic stenosis.
36.	Evans *et al.* (1937), Fig. 15A	Heart failure.
37.	Yater (1938), Fig. 1	Mitral disease.
38.	Conneau *et al.* (1938), Fig. 1	Rheumatic fever.
39.	Katz (1941), Fig. 389	Hypertension.
40.	Wolferth *et al.* (1947), Fig. 2	Angina of effort.
41.	Wolferth *et al.* (1947), Fig. 3	Stokes-Adams syndrome.
42.	Own case	Recent cardiac infarction.
43.	Own case	Cardiac infarction 2 years ago.
44.	Own case	Cardiac infarction 2 months ago.
45.	Own case	Cardiac infarction 8 years ago.
46.	Own case	Cardiac infarction 5 years ago.
47.	Levine (1929), Fig. 58	Recent cardiac infarction.
48.	Appelbaum *et al.* (1934), Fig. 28	Cardiac infarction.
49.	Master *et al.* (1938), Fig. 2	Recent cardiac infarction.
50.	Master *et al.* (1938), Fig. 7A	Recent cardiac infarction.
51.	Master *et al.* (1938), Fig. 8	Recent cardiac infarction.
52.	Katz (1941), Fig. 386C	Recent cardiac infarction.
53.	Wilson *et al.* (1944), Fig. 29	Recent cardiac infarction.
54.	Wilson *et al.* (1944), Fig. 37	Recent cardiac infarction.
55.	Wilson *et al.* (1944), Fig. 38	Recent cardiac infarction.
56.	Goldberger (1947), Fig 79A	Recent cardiac infarction.
57.	Carlotti (1947), Fig. 8A	Recent cardiac infarction.
58.	Curtis Bain *et al.* (1947), Fig. 17A	Recent cardiac infarction.
59.	Curtis Bain *et al.* (1947), Fig. 17B	Recent cardiac infarction.
60.	Soulié *et al.* (1948), Fig. 7	Recent cardiac infarction.
61.	Own case	Cardiac infarction 9 months ago.
62.	Own case	Cardiac infarction some months ago.
63.	Own case	Cardiac infarction some months ago.
64.	Own case	Cardiac infarction 3 weeks ago.
65.	Levine (1929), Fig. 9	Recent cardiac infarction.
66.	Levine (1929), Fig. 58	Recent cardiac infarction.
67.	Evans *et al.* (1937), Fig. 11	Old cardiac infarction.
68.	Master *et al.* (1938), Fig. 1	Old cardiac infarction.
69.	Master *et al.* (1944), Fig. 1B	Recent cardiac infarction.
70.	Wilson *et al.* (1944), Fig. 39	Recent cardiac infarction.
71.	Wolferth *et al.* (1947), Fig. 4	Old cardiac infarction.
72.	Carlotti (1947), Fig. 8B	Recent cardiac infarction.

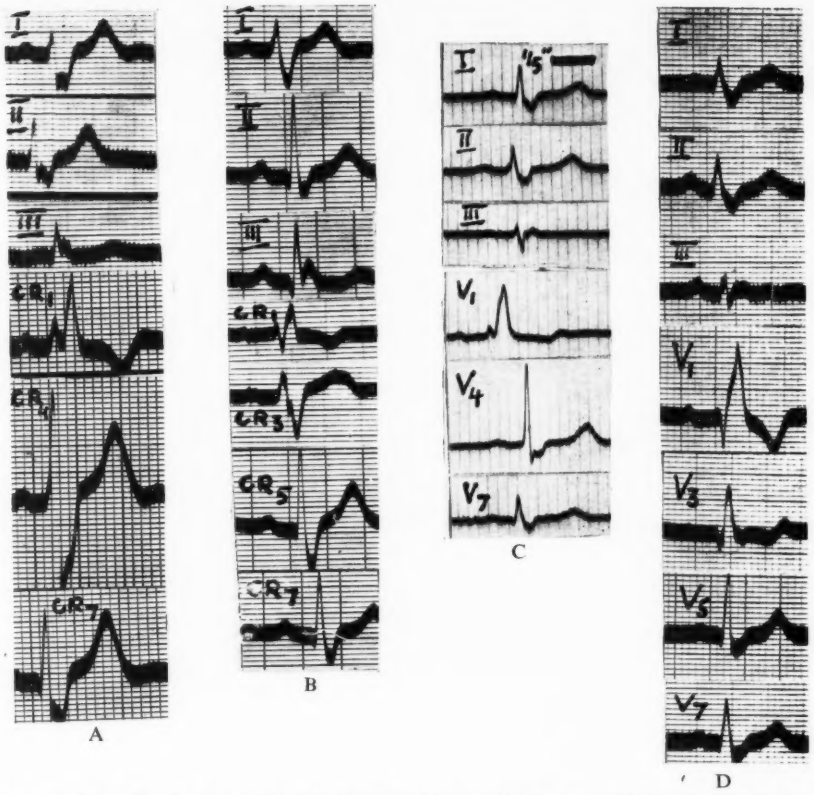
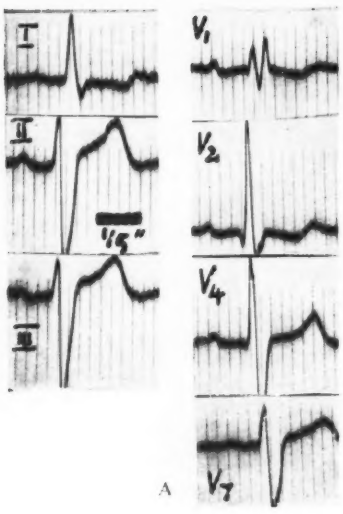


FIG. 1.—Right bundle branch block in healthy subjects. (A) Case 3. (B) Case 7. (C) Case 1. (D) Case 8.

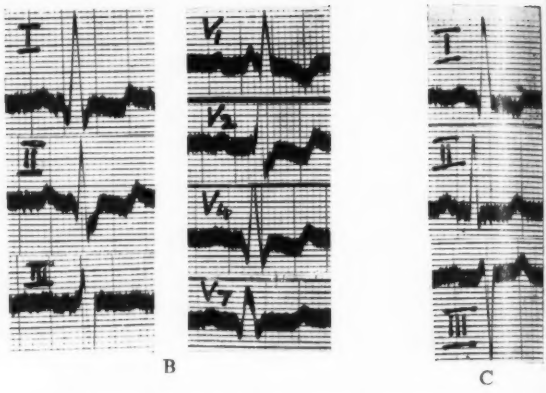
isoelectric line up to the beginning of the T wave, or it started slightly below this line to merge almost immediately into the descending limb of the T wave; the T wave was inverted in all cases except one (Fig. 1C). In left præcordial leads small Q deflections were frequent; an S wave was shown in all records and T was upright. As right branch block caused T inversion in right præcordial leads, so it tended to increase the voltage of T in left præcordial leads.

RIGHT BUNDLE BRANCH BLOCK WITH HEART DISEASE OTHER THAN CARDIAC INFARCTION

This group consisted of 21 cases (see Table, Cases 21–41). In the limb leads. Q I and Q II appeared with the same frequency as in the previous group; the upstroke of R and the beginning of the downstroke of S had generally the same characteristics as those described for healthy subjects, but there were 2 cases in this group with a QRS complex of low voltage. In left ventricular hypertrophy from hypertension or aortic valvular disease (Fig. 2 and 3), the R–T segment often showed depression and a downward slant in lead I, or I and II; the T waves were upright in leads I and II in all cases, though occasionally of low amplitude. Left ventricular preponderance never caused T inversion in right bundle branch block; T inversion shown in normal intraventricular conduction in lead I from left ventricular preponderance was abolished with the onset of right branch block (Fig. 2, B and C), and the amplitude of T waves low in normal conduction was increased in block. Chest leads were taken in 11 cases of this group, and included right præcordial leads in 7. In lead CR1 or V1, a small Q deflection was recorded once (Fig. 2A); the QRS complex showed the bifid R already described, and the T wave remained inverted. In records from patients

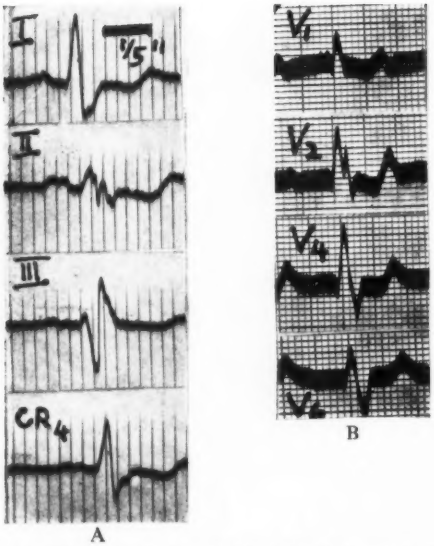


A



B C

FIG. 2.—Right bundle branch block and left ventricular preponderance in patients with aortic stenosis. (A) Case 21. (B) Case 24. (C) Case 24 in normal intraventricular conduction.



A B

FIG. 3.—Right bundle branch block and left ventricular preponderance in patients with severe hypertension. (A) Case 23. (B) Case 26.

with left ventricular hypertrophy, right præcordial leads showed the same type of slanting R–T segment as did lead I, and the T wave was upright in some cases (Fig. 2A and 3B). In apical leads and in leads from the left lateral chest wall, Q deflections were seen in 4 cases, and the deep S of right branch block was present in every tracing. In records with left ventricular preponderance, the R–T segment returned to the isoelectric line, or was even raised above it, as the leads were moved to the left of the chest (Fig. 2 and 3). The T wave remained upright in all left præcordial positions even as far left as V7. None of the patients with left ventricular preponderance were under digitalis treatment.

RIGHT BUNDLE BRANCH BLOCK IN CARDIAC INFARCTION

Nineteen cases of anterior infarction were collected (see Table, Cases 42–60). In the limb leads, Q deflections in leads I and II were neither more frequent nor more conspicuous than in the previous two groups. A low voltage QRS complex was seen eight times; similar low voltage deflections were also recorded with heart disease other than infarction; four cardiograms showed a QRS complex of low voltage and w-shape. The R–T segment in lead I was of coronary type, showing elevation and bowing, in 3 patients; in 8 other cases the R–T segment was elevated in lead I or II, but without bowing, and it was often followed by an upright T wave of normal appearance (Fig. 4, A and B); this

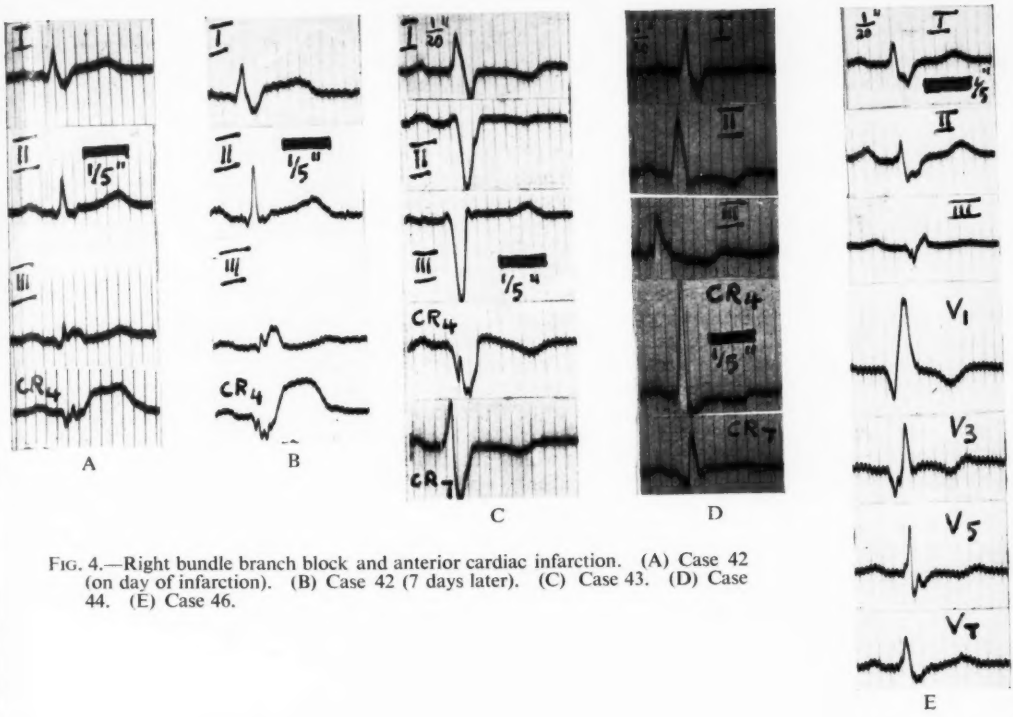


FIG. 4.—Right bundle branch block and anterior cardiac infarction. (A) Case 42 (on day of infarction). (B) Case 42 (7 days later). (C) Case 43. (D) Case 44. (E) Case 46.

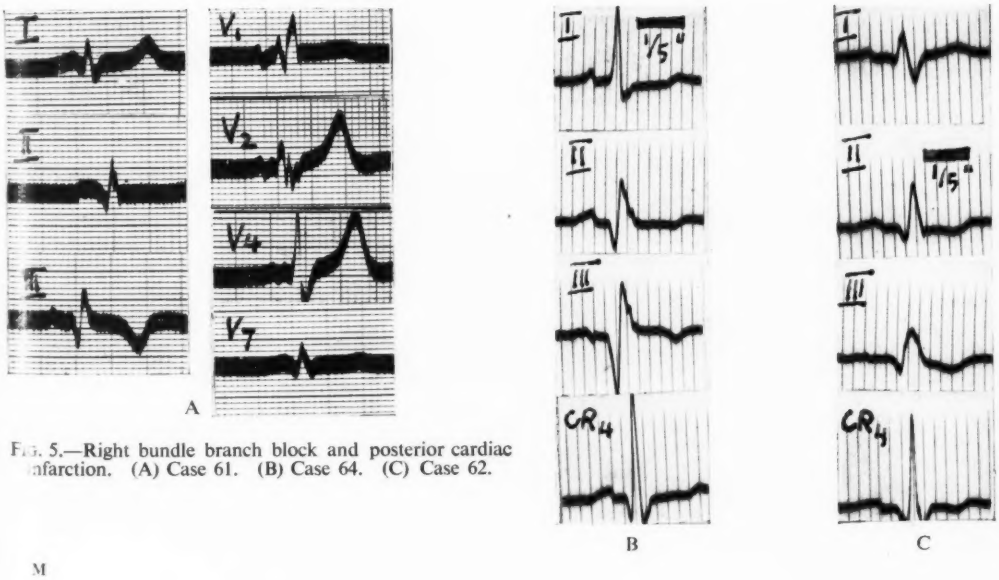


FIG. 5.—Right bundle branch block and posterior cardiac infarction. (A) Case 61. (B) Case 64. (C) Case 62.

form was presumably due to the tendency of the T wave to remain upright in right branch block, and so to prevent the development of typical "coronary" changes. In contrast to the previous two groups, T inversion was a frequent finding in the later stages of infarction; it occurred in lead I in 7 and in lead II in 5 out of 19 cases (Fig. 4, C and D). A number of extrasystoles were seen and examined, but they showed no modifications indicating infarction.

Chest leads were taken in 17 of the 19 cases, and included right præcordial leads in 11. Lead CR1 or V1 indicated the lesion in 9 out of 11 cases in which this lead was taken, by showing a Q deflection followed by a late R with a single peak; recent cases also showed coronary R–T changes. Involvement of this lead seemed to occur more often in right branch block than in normal conduction and to be characteristic of the block; it was not due to a greater frequency of septal infarcts in records with bundle branch block as there was one patient (Case 54) whose cardiogram after infarction showed a characteristic Q in lead V1, only in block and not at other times, in normal conduction. The Q deflection or R–T changes of infarction extended to the apical lead in 5 patients, but were confined to right præcordial leads in 3 (Fig. 4E); in such cases it was important to be certain of the absence of a small initial R wave (compare Fig. 1D and 4E). Lead V1 remained unaffected in two records: in one the lesion was shown in the apical lead (Case 60), and in the other in a lead from the left lateral chest wall (Case 57). Six præcordial cardiograms did not include lead CR1 or V1; in five, changes of infarction appeared in the apical lead (Fig. 4, A to C), and in one in lead CR7 (Fig. 4D).

There were 12 cardiograms in this series showing posterior wall infarction (see Table, Cases 61–72). With the exception of one case of old infarction (Case 71), all tracings exhibited large and significant Q deflections in lead II (Fig. 5); Q I was no more frequent than in any previous group; Q III and inversion of T III, though present in most records, were not significant as they also occurred in right branch block without infarction. T I was upright in all cases, but T II was often abnormal. A coronary type of R–T segment in lead II or III or both was seen in 4 cases. The chest leads analysed in this group included 9 records with apical leads, and 4 with more complete cardiograms. In 3 out of 4 cases, T was upright in lead CR1 or V1 (Fig. 5A); apical leads showed Q deflections in 4 cases and R–T depression in recent posterior infarctions.

Discussion

The preceding analysis has demonstrated a number of modifications of the electrocardiogram of cardiac infarction caused by the simultaneous presence of right bundle branch block. Soon after the common form of right branch block was first recognized in America by Wilson *et al.* (1934) and in England by Evans and Turnbull (1937), the combination of right branch block and infarction was examined by several authors, and especially by Wilson and his collaborators. For the limb leads, Rosenbaum *et al.* (1944) reported the absence of significant Q deflections in lead I in dogs with experimental right branch block and anterior infarction. This observation was confirmed for the human cardiogram in this study. The even R–T elevation, without bowing and with an upright T following it, described here for leads I and II in anterior infarction with right branch block, was not previously discussed, although it was a frequent finding in various published tracings; it was also well shown in cardiograms of dog experiments (Rosenbaum *et al.* (1944), dogs 66 and 71, and Unghváry (1942)). Inversion of T in lead I and II is, of course, a classical sign of anterior infarction, and its significance in records with right branch block had previously been reported by Master, Dack, and Jaffe (1938). The modification of the T wave pattern noted in this study concerned the tendency of of T to remain upright in all cases of right branch block, and especially also in cases with associated left ventricular preponderance. T wave changes, therefore, occurred at a later stage of infarction than in normal conduction, and the significance of T inversion, when present, was enhanced.

Wilson *et al.* (1944) first described the characteristic præcordial cardiogram of anterior infarction, with a Q replacing the initial R of right branch block in lead V1; in the present series, right branch block caused no important modifications of the signs of infarction in apical leads or in leads from the left lateral chest wall, but lead CR1 or V1 indicated the infarct more often than would be expected in normal intraventricular conduction; this seemed independent of the septal extension of the infarcted area expected in many cases of branch block.

No modifications of the limb lead or præcordial cardiogram of posterior infarction have previously been reported. A number of cases here discussed showed upright T waves in chest lead CR1 or V1.

It is now possible to describe, with the help of the preceding analysis, the electrocardiographic signs diagnostic of infarction in the presence of right bundle branch block. The following signs were significant in the limb leads: a w-shaped QRS complex of low voltage and a coronary R–T segment with a coronary T wave in lead I; an even elevation of the R–T segment followed by a normal T wave

and, later, inversion of T in lead I or II or both. The w-shaped QRS complex could not be accepted as conclusive because of the difficulty of distinguishing, in a given case, this form and any other low voltage QRS complex without the w-shape. Inversion of T in lead II was significant except in cases showing the classical and rare form of right branch block; it was, therefore, of no value if R in lead III was conspicuous and taller than R in lead II, and if R in lead I also was of low voltage. Using these signs, the lesion was diagnosed in the limb leads in 16 out of 19 cases of anterior infarction.

This result is not in agreement with previous observations. Wood, Jeffers, and Wolferth (1935) first reported that right branch block may mask the signs of infarction in the limb leads. Master, Dack, and Jaffe (1938) stated that in the presence of right branch block infarction could be diagnosed in two thirds of the cases, and that præcordial leads were more helpful than limb leads; Stokes (1947) quoted this opinion and agreed with it. Wilson *et al.* (1944) reported that in right branch block signs of infarction were rare in the limb leads, but were usually shown in præcordial tracings; this view was also expressed by Goldberger (1947) and Carlotti (1947); Rosenbaum *et al.* (1944) came to the same conclusion for dogs with experimental right branch block and anterior infarction. The explanation for this difference of opinion came from a consideration of the criteria of infarction in the limb leads. In the past, limb lead cardiograms have not been regarded as diagnostic of infarction if the classical signs of infarction were absent. It was here shown that the diagnosis can often be made if the signs include R–T elevation or T inversion, and if the tracings are not expected to show significant Q deflections.

Diagnostic signs of infarction with conspicuous Q deflections and classical R–T changes were seen in the præcordial cardiogram of all 19 cases here reported; in tracings with signs of infarction in apical or left lateral chest leads, the limb leads showed the lesion as well, but right præcordial leads indicated infarction in 3 records in which all other limb and chest leads were negative; in such cases it was important to be certain of the absence of a small initial R wave in lead CR1 or V1.

The diagnosis of posterior infarction could be made from the presence of conspicuous Q deflections in lead II and from coronary R–T changes in lead II and III in all but one case reported here; the præcordial cardiogram showed upright T waves in lead CR1 or V1 in some tracings; this sign had no diagnostic significance as it was not seen in all records of posterior infarction and because it was also present in other conditions, especially in left ventricular preponderance; yet it gave valuable help in the analysis of some tracings. In Case 71 an upright T wave in lead CR1 was the only abnormality in limb and chest leads of a patient known to have had posterior infarction in the past, and a flat T wave in a case of anterior infarction (Case 56) suggested an associated lesion, which was shown to be an old posterior infarct by the limb leads; an upright T in lead V1 of another patient with anterior infarction (Case 60) was explained when left ventricular hypertrophy was found on necropsy.

For the purposes of clinical diagnosis, the limb leads furnished all essential information for the diagnosis of cardiac infarction in most cases; they also indicated left ventricular preponderance. Chest leads CR1 or V1 confirmed the presence of right branch block and disclosed a small number of infarcts not shown by any other lead. The combination of limb leads with lead CR1 or V1 revealed all lesions recorded with more numerous chest leads, and thus satisfied the clinical needs in the 31 cases of right bundle branch block and cardiac infarction reviewed here.

Summary and Conclusion

It was the object of this paper to inquire into the modifications of the electrocardiogram of cardiac infarction in the presence of right bundle branch block, and to examine the criteria necessary for the diagnosis of infarction. The investigation consisted in an analysis of cardiograms with right branch block taken from healthy subjects, from cases with heart disease other than infarction, and from patients with cardiac infarction. Twenty-three patients with right branch block came under personal observation and 49 reported cases were added. Right bundle branch block is shown to cause certain modifications of the cardiographic signs of infarction. In the limb leads of anterior infarction, significant Q deflections are absent in lead I; the R–T segment often assumes a characteristic shape in lead I or II, showing elevation without bowing, and is then followed by a T wave of normal appearance; when T inversion takes place in lead I or II, it more certainly indicates infarction than T inversion in normal conduction, because this change does not occur from left ventricular preponderance in right branch block. In the præcordial cardiogram the infarct is more often shown in CR1 or V1 leads than would be expected in normal conduction; involvement of this lead appears to be independent of the septal extension of the infarct expected in cases of bundle branch block. Right branch block does not change the classical signs of posterior infarction in the limb

leads; in chest leads, the T wave is upright in lead CR1 or V1 in some cases of posterior infarction.

The diagnosis of anterior infarction can be made in the limb leads in many cases if the signs include R–T elevation or T inversion, and if the tracings are not expected to show significant Q deflections; infarction is shown by apical or left lateral chest leads when it is also shown by the limb leads, but lead CR1 or V1 occasionally registers the lesion when all other leads are negative. The diagnosis of posterior infarction is made from the presence of conspicuous Q deflections in lead II and of coronary R–T changes in lead II and III. The præcordial cardiogram may show upright T waves in lead CR1 or V1; although this sign has no diagnostic value, it is helpful in the interpretation of some electrocardiograms.

The limb lead cardiogram in conjunction with lead CR1 or V1 provides the necessary information for the diagnosis of cardiac infarction and right bundle branch block.

I wish to place on record my gratitude to Dr. William Evans for much encouragement and helpful criticism in the study of cardiology and in the preparation of this paper.

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RIGHT VENTRICULAR STENOSIS
(BERNHEIM'S SYNDROME)

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Since this clinical state was first described by Bernheim in 1910, very little has appeared in the English language or in journals easily accessible in this country; or at all in recent years. The recent paper on the subject by Russell and Zohman (1945) gives references many of which we cannot check. A good deal was written in 1930 and 1931, and there were seven papers associated with the name of Mazzei and five with that of Martini. We record these cases to draw attention to a pathological state that appears to explain satisfactorily certain rather curious clinical phenomena.

The essential feature of the syndrome is what appears to be a clinical paradox. There are conspicuous features of apparent failure of the right side of the heart in a patient with a lesion affecting the left side. The explanation for this was offered by Bernheim (1910), who pointed out that the septum of the ventricles bulged into the cavity of the right ventricle, and so prevented it filling. Podestà (1936) has called it dextroventricular stenosis. The progress of the disorder has been divided into two stages by Mazzei (1931). In the first the infundibulum becomes dilated and so allows the flow of blood to be maintained despite the interference with the filling of the ventricle. We have satisfied ourselves by means of casts taken of the cavity of the upper part of the right ventricle in two cases that this is so. In the second phase this adjustment becomes inadequate and the signs of hepatic and jugular engorgement are seen. The pulmonary circulation remains unaffected until near the end, when a final congestion of the lungs may appear.

Case Reports

The clinical and post-mortem findings of our five cases now follow and will be discussed and compared with others reported.

Case 1. A labourer, aged 32, was admitted to hospital in January 1945, complaining of swelling of the legs and feet and of fatigue, but had no shortness of breath. As a child he had attended hospital on account of his heart, and had not been allowed to play games at school. Since leaving school, however, he had been well and had lived an unrestricted life.

The patient was a fat, well-built man. There was no cyanosis. The veins in the neck were not obviously engorged, but the liver was enlarged about one and a half inches below the edge of the ribs. The backs of the legs and the buttocks were swollen. His aspect generally was somewhat pale.

The cardiac dullness extended three-quarters of an inch to the right of the sternum. The apex beat was forcible and situated five and a half inches to the left of the midline in the sixth left interspace. A very loud harsh systolic murmur was best heard three inches from the midline in the fifth left interspace. This murmur was accompanied by a thrill. The murmur was conducted to the mitral and pulmonary areas and to the left axilla, but not particularly upwards. The heart sounds at the apex were quite loud, while those at the base were faint; the pulmonary second sound was louder than the aortic. The pulse was regular, of poor volume, and not anacrotic, with a blood pressure of 100/80. Screening of the heart showed that the right auricle was obviously engorged, the left ventricle was not conspicuously enlarged, and the pulmonary vessels were somewhat prominent. The aorta was normal (Fig. 1).

Barium in the œsophagus showed slight engorgement of the left auricle. The cardiogram showed no axis deviation; the rhythm was normal but the effect of digitalis was apparent in R–T negativity in all three leads (Fig. 2). The diagnosis appeared to have lain between pulmonary stenosis, aortic stenosis, and patent interventricular septum. The site of the murmur, the predominance of right ventricular failure, the absence of an anacrotic pulse

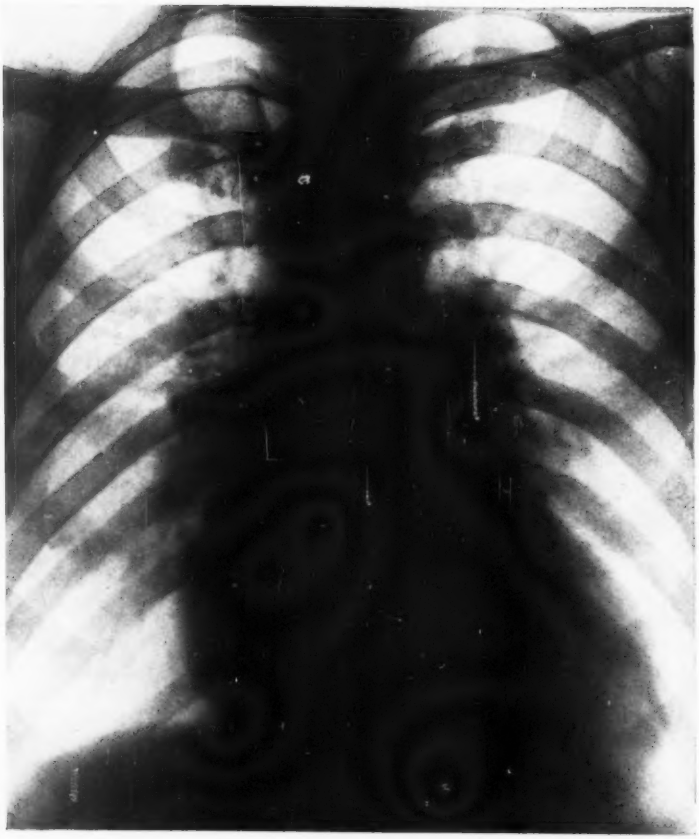

FIG. 1.—Case 1. Some engorgement of pulmonary veins but lungs translucent. Right auricle full: left ventricle enlarged.

and the freedom from dyspnœa, also the equivocal cardiogram, made one decide upon the last, particularly in view of the presence of the lesion in childhood.

At first, treatment for the congestive heart failure was successful. Mercurial diuretics were given freely and he was able to go to a convalescent home. After a month or two he returned with further development of failure. In spite of treatment the anasarca gradually returned and steadily increased. He was never orthopnœic and could lie fairly flat in bed. A pulmonary infarct occurred, and he slowly deteriorated and died.

Autopsy. There was gross anasarca, profuse ascites, and a moderate hydrothorax on both sides. There was no pulmonary œdema and the lungs were not grossly engorged: their dryness was remarked on at the time. The heart was very large—720 g. The left ventricle showed gross hypertrophy, the septum being very thick and bulging extensively into the right ventricle. This was considerably dilated, particularly in the infundibulum. The aortic valves were stenosed as a result of congenital fusion and subsequent calcification. The appearance was that of a bicuspid valve. Calcification was extensive, and in the depths of the anterior cusp there seemed to be traces of a small raphe. The interventricular septum was not patent. The coronary sinus was greatly dilated, perhaps by the high pressure in the right auricle. There was gross passive engorgement of the liver.

Comments. The diagnosis was missed in this case because of the site of the murmur and the absence of confirmatory signs of aortic stenosis

The freedom from embarrassment of the pulmonary circulation, the absence of left axis deviation in the electrocardiogram, and the conspicuous predominance from the first of the results of right side failure made the diagnosis in favour of a patent interventricular septum (Maladie de Roger); for pulmonary stenosis did not appear to be in any way indicated.

Reflection on the findings at autopsy suggested that the clinical manifestations indicated Bernheim's syndrome. The stenosis of the aortic valves should have sooner or later led to failure of the left ventricle, but right ventricular failure was predominant from the first. On Bernheim's theory the encroachment on the cavity of the right ventricle would have been responsible for the early state of the failure of the right side. The absence of engorgement and oedema of the lungs post-mortem was very striking. The absence of left axis deviation agrees with some cases regarded as examples of Bernheim's syndrome, but it seems likely that unipolar limb leads would have shown that the heart was vertical, and consequently the axis deviation due to left ventricular hypertrophy was lacking.

The incorrect diagnosis shows how hard it may be to interpret obvious physical signs, and on reflection it is difficult to see what might have led one to suppose that aortic stenosis was really the cause of the murmur and thrill. Failure of the right ventricle would have been expected to develop early had the ventricular septum been patent.

Case 2. A woman, aged 62, became more and more out of breath on exertion. This symptom was first noted on hills and long walks, and later on climbing stairs. For the last six months she had been breathless on any slight activity. During the winter before coming into hospital she had been confined to bed for two months; there had been several attacks of bronchitis. She was now rather breathless at rest. There was slight swelling of the ankles, but the liver and the external jugular veins were not engorged. The heart rate was 100, with the rhythm regular. The pulse showed alternation and the wave was slightly anacrotic. The blood pressure was 220–210/145. There was thickening of the brachial and retinal arteries. The apex beat was weak and diffuse, extending five inches to the left of the midline, in the fifth intercostal space. In the mitral area a presystolic gallop rhythm was audible.

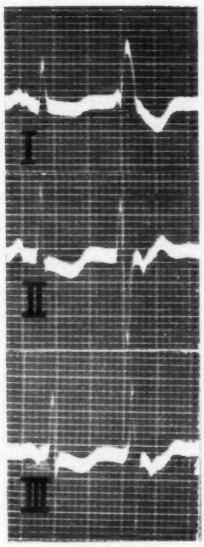

FIG. 2.

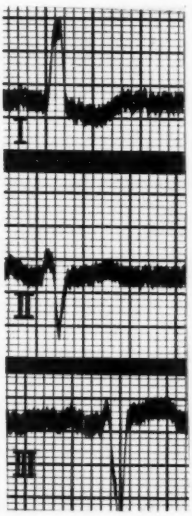

FIG. 3.

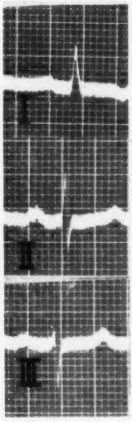

FIG. 4.

FIG. 2.—Case 1. Standard three leads, showing bigeminy and negative T waves, but no left axis deviation.
FIG. 3.—Case 2. Standard three leads, showing left axis deviation.
FIG. 4.—Case 4. Standard three leads, showing left axis deviation.

At the base, in the aortic area a rough systolic murmur could be heard, extending into the arteries of the neck. No thrill was palpable. The aortic second sound was faint; at the bases of the lungs were fine scattered crepitations; the skiagram showed that the pulmonary veins were engorged. The cardiogram (Fig. 3) showed evidence of left ventricular hypertrophy. The diagnosis was failure of the left ventricle due to hypertension and slight aortic stenosis. Failure of the right ventricle was to 22 from 30 a minute. On the evening of the third day she died suddenly.

Autopsy. There was gross œdema and venous congestion. The liver was "nutmeg," weighing 1615 g. The spleen showed severe chronic congestion. The heart weighed 600 g. The left ventricle was enormously hypertrophied, the wall being about 30 mm. thick (Fig. 5). The septum, which was 20 mm. thick, bulged into the cavity of the right ventricle. The aortic valve was stenosed, the cusps

FIG. 5.—Case 2. Transverse section of ventricles looking towards the base. The thickened septum bulges into the right ventricle.

developing. Under treatment she improved and kept fairly well on leaving hospital until six months later, when there was a further attack of bronchitis. This time there was rapid development of œdema and venous congestion. The liver was three inches below the ribs, and there was anasarca to the waist. The jugular veins were prominent half way up the neck. She was rather blue, but not orthopnœic, and could lie fairly flat. A few crepitations were heard in the lungs; the pulmonary second sound was loud. After three days in hospital the respiratory rate fell being fused and calcified. A slight ridge below the free margin suggested a rheumatic infection in the past. The lungs, in contrast to the liver, were free from engorgement, and showed hardly any œdema; the right weighed 550 g. (normal 500 g.), and the left 380 g. (normal 420 g.). Microscopical section showed some engorgement and scattered heart failure cells only. There was but a trace of fluid in the left pleural sac. There was nothing to explain the sudden death except the aortic stenosis.

Comments. This patient, developed gradual

failure of the left ventricle about two years before her death. Œdema of the ankles did not appear until the last six months. The final failure appears to have been precipitated by an attack of bronchitis and was marked by severe venous congestion, which increased rapidly in the last three weeks. Although she was breathless on admission to hospital, this was soon relieved. The important point is the freedom of the lungs from œdema and engorgement post-mortem. It would appear likely that the bulging of the septum into the right ventricle prevented the lungs from being overfilled, although the aortic stenosis and hypertension affecting the left ventricle would have made this a likely finding. It is true that it is common to find failure of the right ventricle following that of the left in such cases as these, and this has always been regarded as a true "back pressure" phenomenon. In this instance it would seem more probable that the cause was different; otherwise the lung would have shown the usual intense engorgement associated with a failing left ventricle. The course of the illness suggests that in the earlier phases the lungs may have been involved but that the development of the stenosis of the right ventricle relieved them.

Case 3. A woman, aged 63, had complained of dyspnœa, gradually increasing in intensity, for the previous four years. During the last six months she had had acute attacks of breathlessness at night and had often been orthopnœic. In the last month the legs had become œdematous, and since then she had been confined to bed. Auricular fibrillation was present, with a heart rate of 70, for she had been taking Guy's pill twice a day for two years. The heart was greatly enlarged to the left, with a heaving apex beat. A rough systolic murmur was heard at the apex. The pulmonary second sound was abnormally loud. Crepitations were heard at the bases of both lungs. Œdema was present halfway up the thighs, and there was a small sacral pad. Ascites was present, and the liver was enlarged two inches below the ribs. The cervical veins were engorged.

A cardiogram showed auricular fibrillation. The limb leads indicated right axis deviation, but the præcordial leads confirmed the clinical evidence of left ventricular hypertrophy.

It was supposed that she had previously had high blood pressure and was now in the later stages of congestive failure with auricular fibrillation. She refused to come into hospital and deteriorated steadily at home; but after a month she had to be admitted. There was now massive œdema of the legs and abdominal wall, and even the hands were somewhat swollen. There were no signs in the lungs and the breathing was easy; she was able to lie quite flat without discomfort.

The cardiogram now showed advanced right axis deviation, with a vertical heart, but the chest leads still indicated hypertrophy of the left ventricle

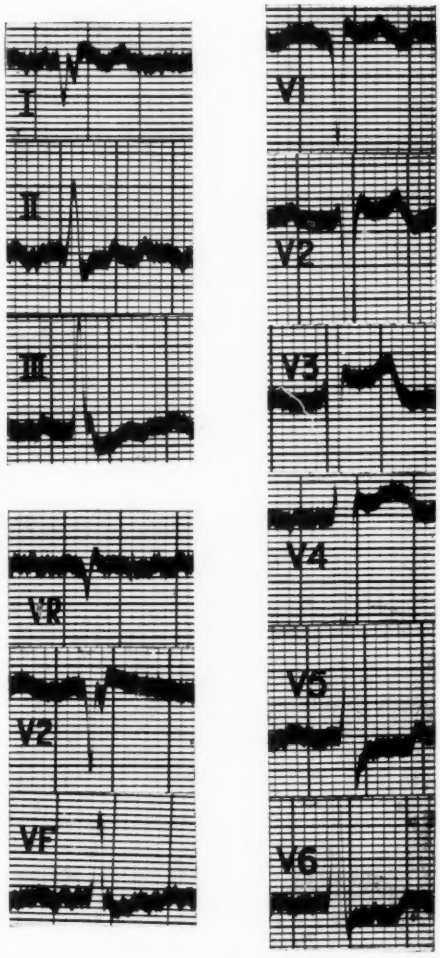

FIG. 6.—Case 3. Standard leads show right axis deviation. Unipolar limb leads show a vertical heart. Unipolar chest leads show hypertrophy of the left ventricle.

(Fig. 6). She slowly got worse, and she died a month later.

The clinical diagnosis of Bernheim's syndrome was suggested on the finding of gross congestive failure in a case of predominantly left-sided disease

together with freedom from embarrassment of the pulmonary circulation. The findings at autopsy confirmed this diagnosis.

Autopsy. Œdema was present in the lower parts of the body. There was no free fluid present in the pleural sacs; the lungs were well ærated throughout and free from œdema. Venous congestion was not conspicuous. The weight of the right lung was 420 g. (normal 500 g.), and of the left lung 390 g. (normal 425 g.).

Section of the lungs showed no evidence of chronic venous congestion. On the whole congestive changes were mild. An early bronchopneumonia was developing in the right lower lobe, and here there appeared œdema in the alveoli and congestion of capillaries.

The heart was greatly enlarged, weighing 810 g. The most striking feature was the concentric hypertrophy of the muscle of the left ventricle. The wall of the right ventricle was also hypertrophied, and the thickness of the interventricular septum was much increased. The thickness of the right ventricle about halfway up was 4 to 5 mm., and that of the left ventricle about the same level was 20 mm. The septum was 20 to 24 mm. in thickness throughout. The cavity of the right ventricle was encroached upon by the bulge of the thickened interventricular septum into it (Fig. 7). There was no apparent dilatation of this chamber. The internal capacity was therefore very considerably diminished. The right auricle was dilated. All valves were normal. There were also numerous areas of atheromatous degeneration in the coronary arteries. The myocardium showed a generalized increase in fibrous tissue which was evenly scattered between the muscle fibres. These fibres themselves showed no degeneration but were definitely hypertrophied. The liver was enlarged (1480 g.) and showed evidence of venous congestion. The kidneys were slightly contracted and granular. There was much free fluid present in the abdomen.

Comments. The conspicuous finding was hypertrophy of the myocardium of the left ventricle, with great increase in thickness of the interventricular septum, which encroached upon the cavity of the right ventricle. The conspicuous venous engorgement of the portal system was in striking contrast with the absence of engorgement in the pulmonary circulation.

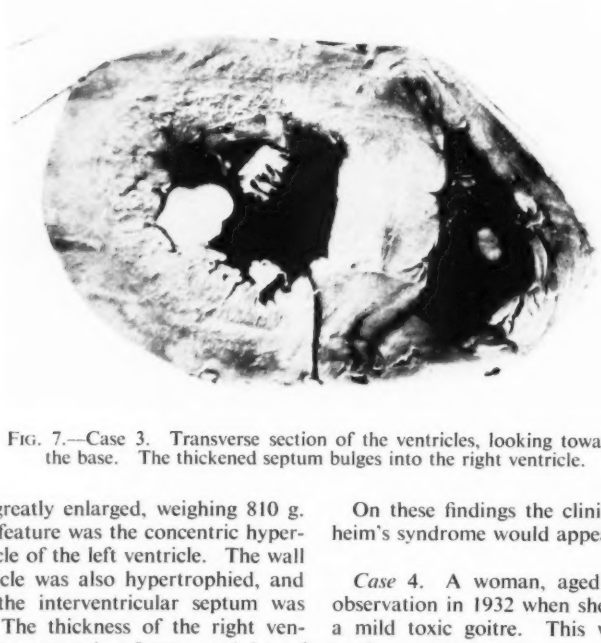

FIG. 7.—Case 3. Transverse section of the ventricles, looking towards the base. The thickened septum bulges into the right ventricle.

On these findings the clinical diagnosis of Bernheim's syndrome would appear to be confirmed.

Case 4. A woman, aged 54, first came under observation in 1932 when she attended hospital for a mild toxic goitre. This was removed and she made a good recovery.

In 1938, when she was 47, she again attended hospital. She had had several severe paroxysms of auricular fibrillation. There had been a swelling of the ankles and pain over the front of the chest. The signs in the heart now showed aortic stenosis.

In 1945, when she was 54, she was again admitted to hospital. During the past seven years she had had attacks of fibrillation from time to time, and in the last year or two she had had a good deal of pain under the sternum on walking. Breathlessness had

not been a conspicuous symptom; there never had been any orthopnœa. There was a good deal of swelling of the ankles and a considerable pad in the sacral area. The veins in the neck were considerably engorged and filled from below up to about four inches above the level of the right auricle when she was sitting nearly upright. The liver was a good deal enlarged. The apex beat reached the sixth space five inches from the midline. There was a typical murmur of aortic stenosis. The aortic second sound was not audible. Auricular fibrillation came and went. There was, however, no satisfactory improvement in the signs of congestive heart failure; digitalis and diuretics were ineffective: gradually the œdema became more general and her condition deteriorated. She was never in any way breathless and she gradually sank and died. The electrocardiogram had shown auricular fibrillation, with left axis deviation.

It was noticeable that this patient, who had evidently had aortic stenosis for a good many years, never complained of shortness of breath. The final phase of heart failure lasted two months and was marked by symptoms of failure of the right ventricle, whereas one would have expected a phase of left ventricular failure.

Autopsy. There was generalized anasarca; a few ounces of fluid were present in each pleural sac but the lungs were remarkably dry, but little engorged and quite free from œdema. The right weighed 510 g. (normal 500 g.) and the left 390 g. (normal 425 g.). The heart weighed 540 g. There was much engorgement of the right auricle and of the great veins. The right ventricle was full; the left ventricle was greatly hypertrophied; and the septum was very thick and bulged prominently into the cavity of the right ventricle. The tricuspid ring was slightly enlarged. The aortic valves were fused, so that a small slit-like opening only was left. The valves were heavily calcified and a ridge of nodular calcium deposit seemed to mark a raphe where the commissure of the two anterior cusps might have been. These were probably congenital bicuspid aortic valves which had become calcified. The stenosis had presumably developed progressively in the course of the last fourteen years.

Comments. In this case it is notable that the pulmonary circulation escaped engorgement right up to the end. One might conclude that this was an example of Bernheim's syndrome in which a terminal "failure" of the right side of the heart occurred, without any indications of failure of the left ventricle. The presence of aortic stenosis would have led one to expect symptoms of left ventricular failure, but in this case again it would appear that engorgement upon the cavity of the right ventricle by the bulging septum precipitated the failure of the right side and protected the lungs.

Case 5. A clerk, aged 58, a tall, thin man, was sent to hospital with severe epistaxis. He was found to have a blood pressure of 220/130, some enlargement of the left ventricle, the apex beat being of a powerful, thrusting character in the sixth intercostal space, four and a half inches from the midline. The peripheral arteries were thick, and the ischæmic, narrow retinal arterioles compressed the veins. There was a trace of albumin in the urine. There was no hepatic or jugular engorgement.

A month later he returned with œdema halfway up the shins, and distended jugular veins, whose fullness increased on compression of the abdomen. The heart rate was 80, the beat regular. A gallop rhythm was audible and with this was associated a duplication of the apex beat, which was easily seen and felt. The aortic second sound was loud, but the pulmonary second sound was not. The cardiogram showed left axis deviation with a negative T I (Fig. 4). He had complained of no dyspnœa at all.

In connection with the last of these indications of embarrassment of the pulmonary circulation it was noted that there were no crepitations at the bases of the lungs, the skiagram (Fig. 8) showed no gross pulmonary engorgement.

After three weeks' treatment all signs of failure had cleared up and the gallop rhythm had disappeared. A few months later he died suddenly at home.

Comment. In this patient signs of congestive failure appeared in the systemic circulation, without the symptoms and pulmonary signs indicating prior failure of the left ventricle. The skiagram was clear; the circulation time (decholin) of only 28 seconds was but little prolonged.

For these reasons it seems correct to class this as an early case of Bernheim's syndrome. Although there was ready response to treatment at first, death occurred a few months later.

Types of Lesion

The underlying lesion in all these cases was either hypertension or aortic stenosis. These cause concentric hypertrophy of the left ventricle. Perusal of Bernheim's original series suggests that most of his patients were hypertensive, but no readings of the blood pressures were made. The same causes were present in Russell's series and those of Mazzei (1931), also in the series of 9 reported by Casaffourth and Superviola (1936). Aortic stenosis was present in Olmer's patient (1933) and in that described by Glushien and Geer (1943). For some reason the

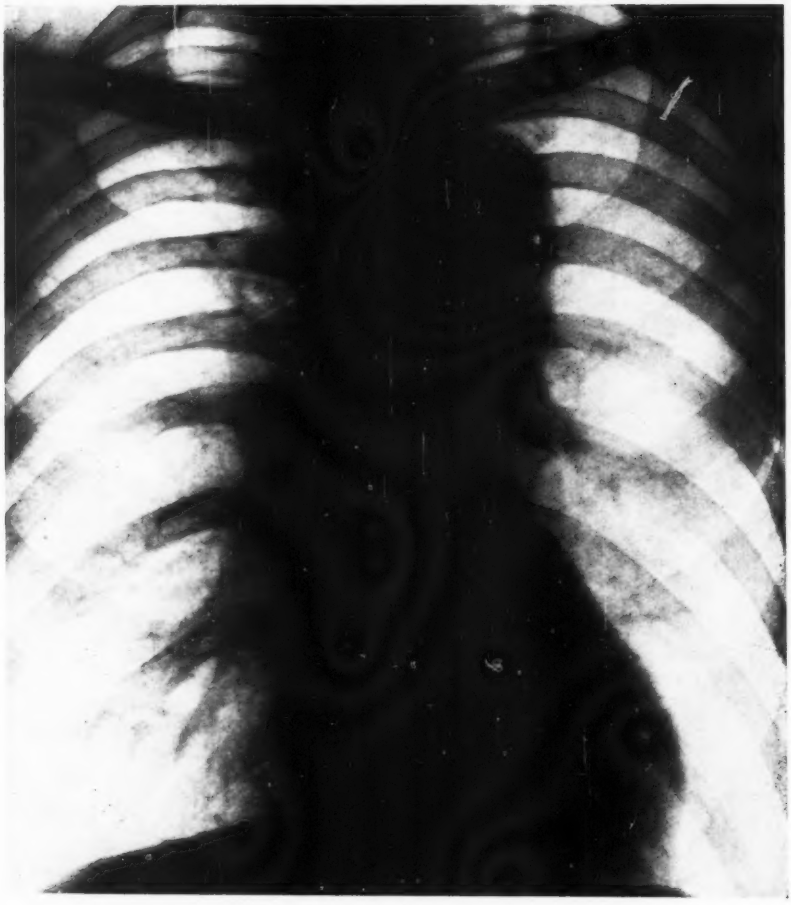

FIG. 8.—Case 5. No indication of pulmonary engorgement. Hypertrophy of left ventricle.

gross dilatation of the left ventricle caused by free aortic incompetence does not lead to right ventricular stenosis. Possibly the explanation is that the septum does not become sufficiently thick.

Appearances in the skiagram. It has been claimed that distension of the right auricle is a feature (Glushein and Geer, 1943). Two of our cases suggested this, but in Case 5, admittedly in an early stage (Fig. 8), it is not present. The important point is the absence of engorgement of the fields of the lungs, although there may be slight overfilling of the pulmonary veins. We agree with other observers that the important triad is large left ventricle, clear lung fields, and full right auricle.

POST-MORTEM FINDINGS

The encroachment of the septum upon the cavity of the right ventricle is very obvious when the ventricles are cut across at right angles to the long axis of the heart, midway between the apex and the base. This finding is likely to be missed when the ventricles are opened by V-shaped cuts, one down the outer border and the other upwards to the pulmonary artery (or aorta), as is usually done. The transverse cut should really be made first of all in any post-mortem examination of the heart; this procedure might prevent the septal bulging from being overlooked, as we fancy it easily may be if the other procedure is followed.

Photographs of the transversely divided ventricles are apt to be unsatisfactory, as by the time the heart has been opened the relative positions of the walls of the ventricles is lost: and plugging with cotton wool and fixing in formalin easily spoils the true shape. We attempted to make a cast, in two cases, of the upper part of the right ventricle and infundibulum. It is almost impossible to get a good photograph of this, but we were satisfied that this part, the outflow tract, remains tubular in form, leading up to the pulmonary artery, thus giving an outlet relieving the constriction below, as suggested by Russell and Zohman (1945) and earlier by Mazzei (1931).

The Lungs

The dryness of the lungs at autopsy, free from engorgement and œdema, as shown by their normal weight, is very striking. In all four cases this was a notable feature. This agrees, of course, with the relatively rapid circulation rate. Other observers have noted (e.g. Bernheim in five of his ten cases) infarcts in the lungs: whether these are due to emboli or thromboses is not clear.

Cooke and White (1941) point out that in tricuspid stenosis there is conspicuous jugular engorgement with freedom from pulmonary congestion, so that the patient may be able to lie flat and even walk about with but little distress. Constriction of the pericardial sac presents the same apparent paradox, as Glushien and Geer (1943) have noted. The syndrome of Bernheim, by reason of the right ventricular stenosis, presents a third example of this curious combination of signs; it amounts, in fact, as Fishberg has stated (1940), to "a virtual tricuspid stenosis."

Other Findings

The circulation rate. In two of our patients (Cases 1 and 5) in which the observation was made the circulation rate was not grossly slowed, the arm to tongue time in both being 28 seconds. This is quite unlike the result one would expect in ordinary congestive failure, when it should be well over 40 seconds. Similar observations were made by Russell and Zohman (1945). As slowing chiefly occurs in the pulmonary circulation, the results indicate the relative freedom of the lungs from embarrassment.

The cardiogram. It will be noted that three of the curves in these cases of left ventricular hypertrophy show the expected left axis deviation in the three standard limb leads. In one this feature is absent and in another there is actually right axis deviation. There has been a good deal of comment on this absence of the curve of left axis deviation, and it has been suggested that the bulging of the septum of the ventricles to the right may be the cause of these curves by affecting the electrical axis (Russell, Zohman, 1945). It seems to us that the explanation is to be found in the unipolar limb leads, which show that the heart is actually vertically placed in the chest, and so the presence of the hypertrophy of the left ventricle cannot cause left axis deviation. This is not an uncommon finding in certain cases of left ventricular hypertrophy; quite apart from the presence of septal displacement to the right. We have recently noted three patients with aortic stenosis, two congenital in youths, and the third of the fibrocalcareous acquired type in an elderly man, who showed no left axis deviation, but with unipolar limb leads showing the heart to be vertical.

We therefore conclude that there are no cardiographic changes peculiar to Bernheim's syndrome.

Further investigation. It seems likely that two lines of investigation in such cases as these will be fruitful. It would be interesting to know what pressures may be revealed in the pulmonary artery by intracardiac catheterization. Angiocardiography should show the peculiar shape of the cavity of the right ventricle.

Treatment

In these cases the question of venesection may arise. It would seem inadvisable to bleed in these circumstances, as a raised venous pressure is no doubt beneficial. Anything that reduces the venous pressure might do harm; and this perhaps applies to such drugs as digitalis and cardophylin.

Summary

Right ventricular stenosis (Bernheim's syndrome) may be found in patients with hypertension or aortic stenosis.

The symptoms suggesting failure of the right ventricle come on early, and may be transient at first. The pulmonary circulation remains free from embarrassment, and the patient is free from orthopnœa, even to the end.

The clinical diagnosis is confirmed at autopsy by the state of the lungs and transverse section of the ventricles.

There are no cardiographic changes peculiar to the condition.

Our thanks are due to Professor Magnus for Fig. 5, and Dr. J. V. Wilson for Fig. 7.

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MITRAL STENOSIS IN LATER LIFE

BY

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Rheumatic heart disease with mitral stenosis has been regarded as a disease that produces symptoms in adolescence or early adult life, and death before the age of forty in the majority. It is admitted that occasionally the first symptoms will appear at the time of the menopause and that if the lesion is accompanied by hypertension, life may be prolonged beyond this period (Levine, 1945). The few clinical observations that have been made on the subject, put the incidence of mitral stenosis beyond the age 50, at from 6 to 10 per cent (Coombs, 1924; Brenner, 1934; Hedley, 1940) (see Table I). Levine and

TABLE I

MITRAL STENOSIS (OR RHEUMATIC HEART DISEASE)
(Proportion of cases over 50 years of age)

Clinical cases Author	Necropsy cases Author
1. Coombs (1924) 6%	1. Coombs (1924) 9%
2. Brenner (1934) 10%	2. Cabot (1926) 33%
3. Hedley (1940) 7%	3. Cookson (1930) 12% (M.S. and A.F. over age 48)
	4. Brenner (1934) 20%
	5. Hedley (1940) 11%
	6. White and Bland (1941) (5 cases, aged 73–85)
	7. Gelfman (1943) 33%
	8. Zeman (1945) (10 patients aged 60–74 with M.S. and subacute bacterial endocarditis)
	9. Karsner and Koletsky (1947) (48 with M.S. and aortic calcification in elderly patients)

Kauver (1941–42) were able to collect from the records of a hospital and a private practice over about a quarter of a century, 28 patients over 50 years of age with mitral stenosis and angina pectoris. But more information is available from post-mortem studies than from clinical reports on the question of mitral stenosis in the elderly. These reports give the percentage of patients dying with mitral stenosis at age 50 or more as from 9 to 33 per cent, figures that are so much higher than those for the clinical incidence as to suggest that the diagnosis is often missed during life (Coombs, 1924; Cabot, 1926; Cookson, 1930; Brenner, 1934; Hedley, 1940; White and Bland, 1941; Gelfman, 1943; Zeman, 1945; Karsner and Koletsky, 1947) (Table I).

The present investigation concerns 37 patients with mitral stenosis and one with mitral incompetence, ranging in age from 51 to 77 years. There were only 9 men, giving a sex ratio of more than 3 females to 1 male, which is higher than that for all cases of mitral stenosis. (Table II.) The

TABLE II

THIRTY-EIGHT PATIENTS WITH MITRAL DISEASE OVER FIFTY

Age	Number
51–60	26 (18 F. 8 M.)
61–70	8 (7 F. 1 M.)
71–77	4 (4 F. 0 M.)

predominance of women increased in each successive decade so that all patients over 70 were women. A greater number was seen in private than in hospital practice which is the reverse of the distribution for younger age groups. There was a clear history of rheumatic fever or chorea in 15, and in a further 8, a valve lesion or heart disease had been diagnosed in childhood or early adult life. In 5, the first known attack of rheumatic fever occurred at the age of 34 or later, and recurrent attacks after the age of 30 were mentioned in 2 others.

All had lived actively and, though some admitted that they had never been capable of strenuous exertion because of shortness of breath, the general capacity for work was at least average. Of 23

women who were married, 18 had had children, in one case, five.

Initial Symptoms

When first seen, 32 of the 38 cases had established auricular fibrillation; 2 came under observation with paroxysmal tachycardia, 1 with paroxysmal fibrillation, and only 3 had normal rhythm. In one of these last, fibrillation set in after a few weeks. In 8 cases a sudden onset of symptoms, mainly palpitation and dyspnœa, was known to coincide with the occurrence of fibrillation or was strongly suggested by the history. In 3, the first symptoms were due to cerebral embolism, in one to pulmonary infarction and in one to ischæmia of the legs. Congestive failure was present in about one third of all cases when they were first seen, and this nearly always responded to appropriate treatment. In a further third, systemic congestion developed during the period of observation and treatment which was generally a matter of years. Failure under these conditions was resistant to treatment though some survived in a state of invalidism for months or years. Systemic congestion with normal rhythm was observed in one patient only. Two patients who died from the effects of left auricular thrombosis never showed signs of systemic congestion. They had auricular fibrillation and symptoms for six months and four years respectively.

Diagnosis

The diagnosis of mitral stenosis in the patients in this series, was made on the presence of a rough, low-pitched diastolic murmur heard best at the apex beat or occasionally a short distance from it, as for example in the intercostal space above the apex. The murmur begins very soon after the second sound, is decrescendo, dying away as a rule before the first sound, though rarely reaching it. In this last event an impression of a crescendo presystolic murmur is possible even with auricular fibrillation. The diastolic murmur may be so rough as to be unmistakably that of mitral stenosis, but more often it is softer, yet not of the blowing quality characteristic of the aortic diastolic murmur. Nevertheless, when aortic incompetence was present, as it was in ten patients, mitral stenosis was not considered proved by a low pitched apical murmur, unless characteristic X-ray changes of mitral stenosis were present. The diastolic murmur of mitral stenosis in the elderly requires careful and sometimes lengthy auscultation for its detection, and it may be quite inaudible in any but the long diastoles. Mitral stenosis may of course exist when the only murmur is systolic. There were two cases of this type, in which the diagnosis was established post-mortem, but apart from these a mitral diastolic murmur had been heard in all, though not necessarily on every occasion the patient was examined. Subsidiary signs of mitral stenosis were usually present; in order of frequency these were a sharp first sound, a third heart sound, accentuated pulmonary second sound, and flushed or cyanotic appearance of the malar eminence.

Hypertension

Levine (1945) remarked on the frequency of arterial hypertension in older patients with mitral stenosis, and indeed ascribed their longivity to this complication. It is not easy, however, to determine the frequency of hypertension in these cases as the blood pressure is difficult to estimate. This is, first, because of the almost invariable presence of auricular fibrillation and second because there is sometimes aortic incompetence, both of which conditions tend to give high readings for the systolic and lower readings for the diastolic pressure. In the present cases a minimum diastolic pressure of 100 mm. or more was found in 19 (50 per cent). On this criterion of hypertension, it was present in one half of all patients. Taking a systolic pressure of 190 mm. or more as a criterion of hypertension, it was present in 12 (31 per cent) of cases. With either of these standards the incidence of arterial hypertension must be regarded as high in mitral stenosis in the elderly.

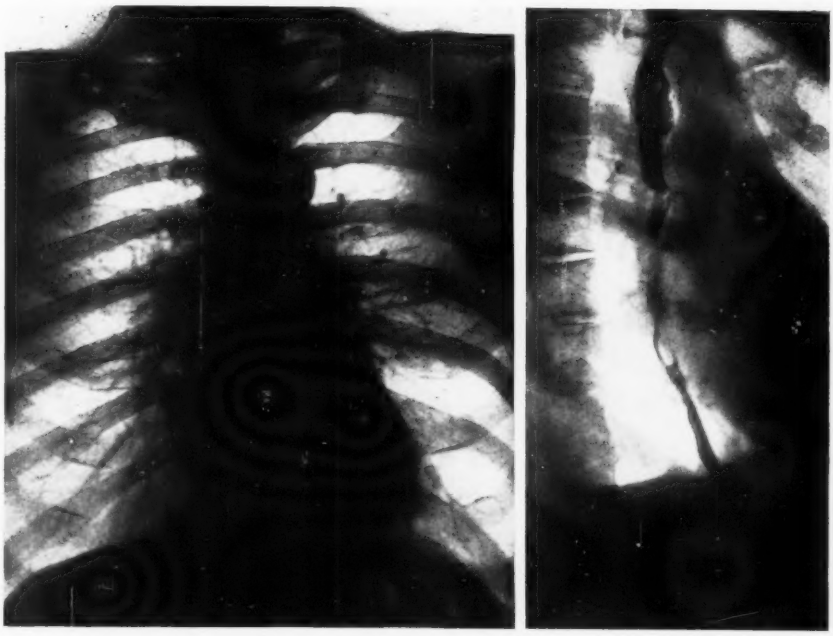
Yet in spite of this high proportion with hypertension and the advanced average age of the subjects, characteristic anginal pain was experienced by one patient only. This was a woman of 71 with mitral stenosis, aortic incompetence, and auricular fibrillation. Her blood pressure was 160/100 mm. She described typical retrosternal pain in walking up steep hills. She had never shown any signs of systemic congestion. In a further two patients, however, pain occurred which may be regarded as anginal. This was in 2 men who died of left auricular thrombosis and who complained shortly before death of pain in the lower chest and epigastrium.

Radiological Appearances

The accepted X-ray cardiac changes in mitral stenosis are an increase in the left middle arc so that the usual slight concavity in this region comes into line with the left lower contour and the shadow of the aortic knuckle, or there is a convexity in this region which may be single or double. The vascular pedicle is narrow and the aortic knob is small or non-existent. Displacement of the œsophagus

backward and to the right reveals an enlargement of the left auricle. At first these changes are not accompanied by any alteration in the shape of the left lower contour or of the right contour, but as time passes these also become more or less prominent, and on the right border may be seen a double contour produced by the two auricles.

Radioscopy and radiography were carried out in all cases except five, and in three of these five, necropsy was done. A cardiac silhouette that could be regarded as within normal limits was seen only once. (Fig. 1A and 1B.) This was in a man aged 58 whose only known attack of rheumatic fever occurred when he was 45: auricular fibrillation was present but symptoms were very mild. Slight general cardiac enlargement with increase in the left middle arc was also unusual. An example is shown in Fig. 2 taken from a man of 53 who had normal blood pressure and whose heart at necropsy weighed 325 g., with moderate right ventricular hypertrophy, the left ventricle being normal. The onset of auricular fibrillation causes a rapid increase in the size of the heart even in the absence of systemic congestion. This is shown in Fig. 3 and 4 from a woman aged 51. The first of these radiographs was taken while normal rhythm was present and the second a few weeks later after auricular fibrillation had developed; it shows some increase in the right, left lower and left middle contours of the heart. Increase in the size of the left auricle sufficient to bring it to the right heart border was seen in five cases, although in the first oblique position they usually showed only moderate displacement of the œsophagus. Deviation of the œsophagus to the left and backward instead of to the right and backward was met with in two patients. (Fig. 5, 6, 7, and 8.) This is well seen on screening after barium swallow in the second oblique position or in the face position, but in the first position the course of the œsophagus may appear almost normal. The narrow vascular pedicle and inconspicuous aortic knob characteristic of the X-ray picture in young subjects with mitral stenosis was never seen. On the contrary the knob was always visible and often prominent, though this prominence tends to be minimized by the increased middle arc below it.



A B

FIG. 1.—Man, aged 58; an Army officer on active service until age 56; only known attack of rheumatic fever at age 45; mitral stenosis, auricular fibrillation. (A) Postero-anterior radiograph shows heart of normal size and shape, or possibly minimal increase in left middle arc. (B) Right (I) oblique view shows no enlargement of left auricle.

When the left lower contour is also well out, the result is a contour which bears no resemblance to the typical silhouette of mitral stenosis.

A well marked aortic impression on the œsophagus and the opacities of aortic calcification are also common features of the X-ray picture of mitral stenosis in the elderly. Fig. 9 and 10 represent the characteristic features of mitral stenosis combined with great cardiac enlargement in three women aged 71, 74, and 77 respectively (see pp. 162–163).

Cause of Death

Sixteen of the 38 patients died after an average duration of symptoms of 4½ years, the average age at death being 62 years. Twelve died with systemic congestion complicated in two by cerebral vascular lesions, acute rheumatism in one and pulmonary infarction in one. Two died from acute pulmonary congestion. In four of the sixteen cases necropsy was done. Two of these showed thrombosis of the left auricle, the thrombus extending on to the mitral valve and further obstructing it. The first, a man aged 60, had some abdominal and leg pains, then developed a sudden painful paraplegia, with pain also in the chest and left arm and delirium. There was a complete absence of arterial pulsation in the legs which quickly became gangrenous. Death occurred on the 19th day. Necropsy showed an extreme mitral stenosis, the orifice just admitting the tip of the little finger. A large soft vegetation measuring 3 cm.×2 cm. was situated on the left auricular wall extending on to the aortic cusp of the mitral valve and encroaching on its opening (Fig. 11). Just above the bifurcation of the aorta there was a small dissecting aneurysm of its wall with leakage of a little blood into the retro-peritoneal tissues (Fig. 12, page 164).

The second, a man of 53, had lower chest or epigastric pain with symptoms of cerebral anoxæmia a few hours before death. Necropsy again showed extreme mitral stenosis with a loose fresh-looking

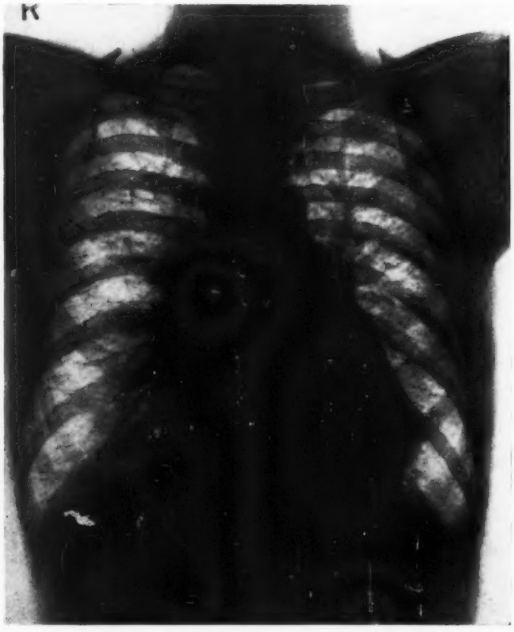


Fig. 2.—Man, aged 53. Mitral stenosis and auricular fibrillation; postero-anterior radiograph shows prominence of pulmonary artery and of conus separately, but no prominence of other cardiac contours; aortic knob prominent. *Necropsy:* severely stenosed calcified mitral valve with fresh thrombus on its auricular aspect; left ventricle not enlarged; heart weight, 325 g.

clot blocking the orifice. Rough calcified plaques almost encircled the base of the valve. Neither of these two patients with severe mitral obstruction had shown signs of systemic congestion. Their chest pain near the end may have been cardiac in origin and due to auricular thrombosis as suggested by Evans and Benson (1948). In a third case, a woman of 62, necropsy showed pure mitral incompetence. The clinical diagnosis had been hypertension and congestive failure, though there was a history of rheumatic fever in childhood, and valve disease had been diagnosed at age 7. The layers of the pericardium were densely adherent throughout; the mitral cusps were thick and opaque with rounded margins and the orifice easily admitted four fingers. The left auricle was greatly enlarged with a capacity of over 500 ml. (Fig. 13.) A fourth necropsy on a woman of 59 who died in congestive failure, shows a heart weighing 750 g. with severe mitral stenosis and moderate aortic and tricuspid stenosis.

TABLE III

CAUSE OF DEATH IN SIXTEEN PATIENTS

(*Average age at death* 62 *years*)

Systemic congestive failure	12
(Acute rheumatism, 1)	
(Cerebral vascular lesion, 2)	
(Pulmonary infarction, 1)	
Acute pulmonary œdema	2
L.A. thrombus obstructing mitral orifice	2

DISCUSSION

The onset of auricular fibrillation appears to mark a turning point in the life history of patients who have suffered little or no handicap from mitral stenosis with normal rhythm up to middle or late life. Symptoms are rarely sufficient to make the patient seek advice till an arrhythmia develops, nearly always fibrillation, but occasionally it is auricular tachycardia that is responsible for the breakdown. Only three patients had normal rhythm when they first came under observation. Once fibrillation has become installed the patients' activities are more or less curtailed and their lives must thereafter be lived in a state varying from slight

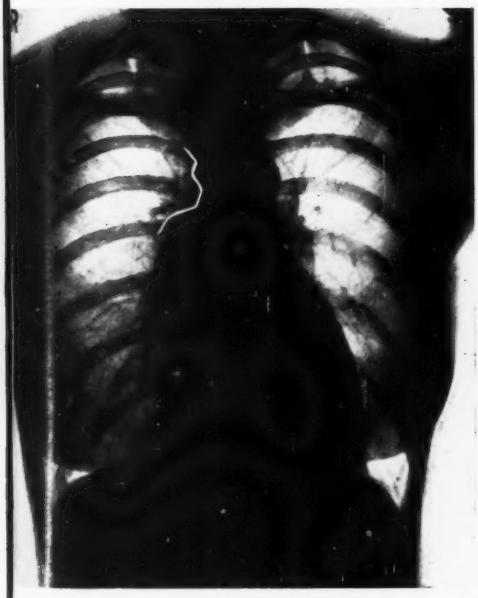


FIG. 3.—Woman, aged 51. Mitral stenosis, normal rhythm; radiograph shows some increase in left middle arc only.

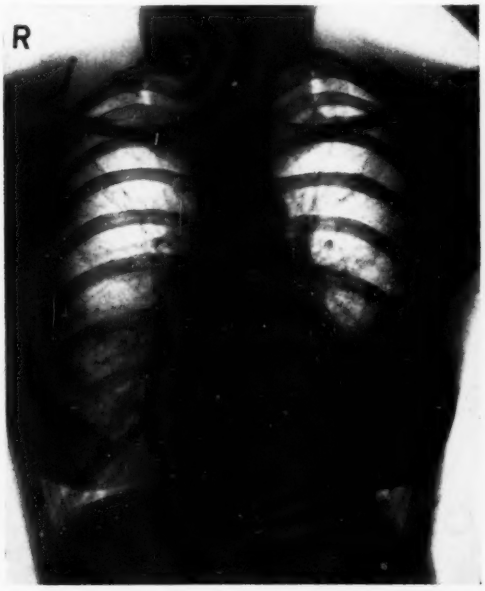


FIG. 4.—Same case as Fig. 3. Radiograph taken a few weeks later, after onset of auricular fibrillation, now shows greater prominence of left middle arc and some increase in left lower and right contours, although there were no clinical signs of venous congestion.

to severe invalidism. At the first appearance of fibrillation with a rapid ventricle or of paroxysmal tachycardia, systemic congestion may occur but this is usually amenable to treatment after which there may be a long period of fair comfort. In some cases systemic congestion is delayed for years after the onset of fibrillation but when it does eventually occur it is resistant to treatment. Life may nevertheless be prolonged for years in a condition of advanced failure. The duration of symptoms that were more or less incapacitating averaged 4½ years for the whole series with extreme for a few weeks to 25 years. The commonest cause of death was systemic congestion with pulmonary infarction and cerebral vascular lesions as occasional contributory causes, but in two patients, one with severe hypertension, it was left ventricular failure. Two patients who died from left auricular thrombosis are of interest, in that neither had shown systemic congestion in spite of severe mitral stenosis.

The radiographic appearance of the heart in mitral stenosis of later life shows some differences from the typical picture in young subjects with this valve lesion; the heart is usually much enlarged and the aortic knob is not small; often it is prominent and shows calcification of its wall. Barium swallow sometimes shows a conspicuous aortic impression. Left auricular enlargement was revealed by the œsophagogram, yet in cases where the enlargement was great, as indicated by its extension to form part of the right cardiac border, œsophageal displacement in the first oblique position was only moderate. In two the œsophagus was displaced considerably to the left and backward. Displacement in this direction can be seen only by screening in the second oblique or face positions; it is very rare in mitral stenosis in young subjects.

Although a valve lesion or heart disease had been diagnosed in 8 of the patients when they were young, only 3 of the 38 cases described were referred with the diagnosis of mitral stenosis and it seems probable that many elderly patients with this valve lesion are not recognized as such. The mitral diastolic murmur often needs careful auscultation for its

FIG. 5.—Woman, aged 58. Mitral stenosis, auricular fibrillation, hypertension (260/110), congestive failure; working as domestic servant until a few days previously. Postero-anterior radiograph shows great cardiac enlargement, straight left border; œsophagus seen through heart shadow, is deviated to left. Right pleural effusion.

FIG. 6.—Same case as Fig. 5. Left (II) oblique view; left-auricular enlargement shown by displaced œsophagus.

FIG. 7.—Same case as Fig. 5 and 6. Right (I) oblique view; œsophagus not displaced by left auricle but the shadow of the L.A. can be seen extending backward across the spine. Calcification of aortic arch.

detection when auricular fibrillation is present and the site of audibility may be very localized. Moreover there is a general reluctance to diagnose rheumatic heart disease in the elderly, which makes it more unlikely that the characteristic signs will be found. Hence a diagnosis of arteriosclerosis or hypertension—which are also frequently present—is made. There is little reason to think that an atheromatous lesion with calcification was responsible for any of the present cases, and it is not established that such a lesion can produce mitral obstruction of clinical importance. In about 60 per cent there was a clear history of rheumatic fever or of a cardiac lesion having been found in early life. The absence or mildness of symptoms in the present group until late in life, is perhaps to be accounted for by a relatively slight myocardial injury; in a few perhaps by the first attack of rheumatic fever occurring at a relatively advanced age. It is not due merely to the slightness of the valvular stenosis, as indicated by extreme narrowing of the mitral valve in two patients who came to autopsy, neither of whom had ever had congestive failure. White and Bland (1941) describe similar severe latent mitral stenosis in three patients aged 73 or more. It is evident that as with some other obstructions to the main vascular channels, such as severe aortic stenosis and aortic coarctation, an advanced lesion of this valve is compatible with good health over very long periods of time.

Summary

Thirty-eight patients, aged 51 to 77, with mitral stenosis (including one with pure mitral incompetence) are described. The valve lesion was believed to be rheumatic in all cases. Sixty per cent gave a clear history of rheumatic fever, or of a cardiac lesion having been discovered in early life. In a few cases the first known attack of rheumatic fever occurred in the fourth decade or later. Physical activity had been little, or not at all, restricted in the past and the appearance of symptoms calling for medical care coincided nearly always with the onset of auricular fibrillation, though occasionally with paroxysmal tachycardia. There was arterial hyper-

FIG. 8.—Woman, aged 66. Mitral stenosis, auricular fibrillation, hypertension (245/150) and congestive failure. Great general enlargement of the heart; very slight convexity of left middle arc; œsophagus pushed to left by large left auricle; deep aortic impression on œsophagus; calcification of aortic knob.

tension in a high proportion of the patients. The X-ray picture differs in several respects from that of younger subjects with mitral stenosis. Initial symptoms, occurrence of congestive failure and cause of death are discussed. It is suggested that mitral stenosis is not rare in patients over 50 years of age and that careful auscultation of elderly subjects with auricular fibrillation will reveal this valve lesion, where formerly it was unsuspected.

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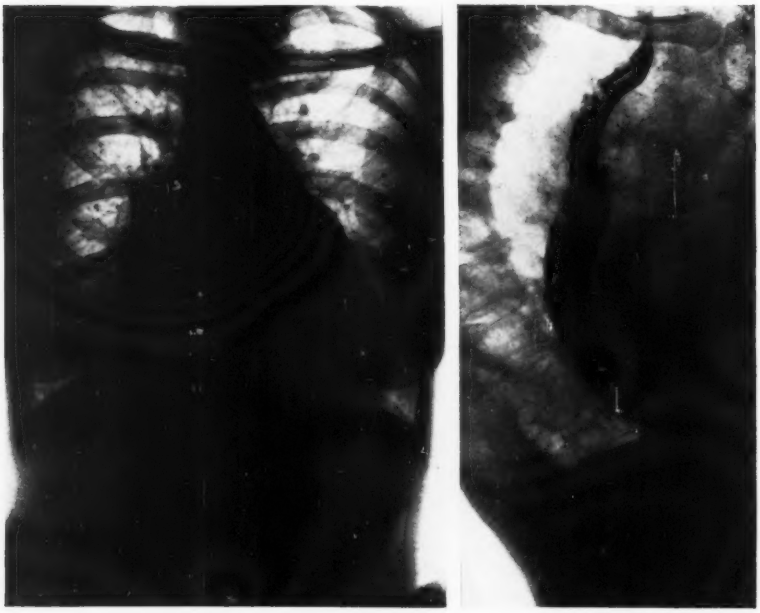

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FIG. 9.—Woman, aged 71. Mitral stenosis, slight aortic incompetence, auricular fibrillation; no signs of systemic congestion; B.P. 160/100. (A) Postero-anterior view shows considerable cardiac enlargement; small convexities on upper part of left profile. (B) Right (I) oblique view shows œsophageal displacement by left auricle.

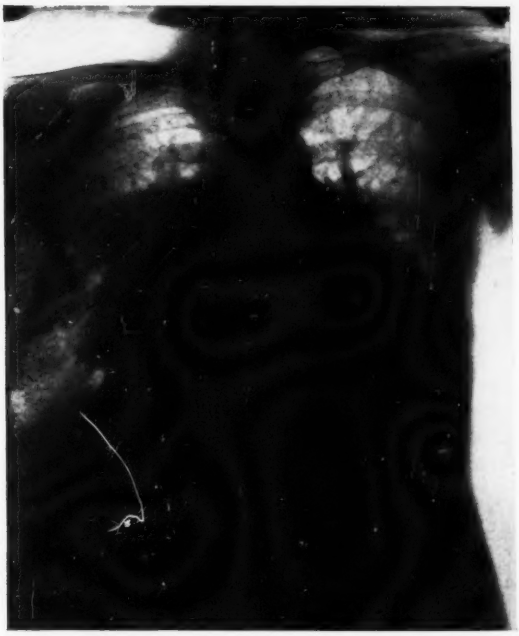

FIG. 10.—Woman, aged 77. Mitral stenosis, aortic incompetence, auricular fibrillation, congestive failure. Postero-anterior view shows very great cardiac enlargement of crinoline shape with a relatively narrow waist above formed by the vascular pedicle; left middle arc very prominent; pulmonary opacities suggesting hæmosiderosis.

FIG. 11.—Heart from man aged 60. Mitral stenosis, auricular fibrillation; no systemic congestion; death from gangrene of the legs. Photograph shows opened left auricle to the left of the specimen; and a thrombus measuring 3 cm. × 2 cm. on the wall of the auricle which extends on to the aortic cusp of the mitral valve and further obstructs its orifice. Mitral opening just admitted tip of little finger.

FIG. 12.—Abdominal aorta from same patient as Fig. 11. Shows perforated atheromatous ulcer just above the bifurcation.

FIG. 13.—Heart from woman aged 62. History of rheumatic fever in childhood. Clinical diagnosis hypertension (215/115) auricular fibrillation, congestive failure. Necropsy showed adherent pericardium and as shown in photograph, large left auricle with a capacity of 500 ml.; thickened opaque mitral cusps with rounded margins, the orifice admitting four fingers. This was regarded as rheumatic mitral incompetence without stenosis.

CIRCULATION TIMES IN CONGENITAL HEART DISEASE

BY

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Since the introduction of surgical treatment for congenital heart disease, which was mainly of academic interest before, it has become important to diagnose as far as is possible the anatomical and dynamic abnormalities present in each case. This need has placed a premium upon any method, more especially if simple and safe, that may help to elucidate the problem. The purpose of this communication is to report briefly the results obtained from circulation time estimations in congenital heart disease, and to discuss what value this simple test possesses.

References to circulation times are numerous, and it is not intended to review these extensively. Blumgart, Weiss, and others first perfected a technique and studied circulation times extensively, reporting their results in 1927 and 1928. Their method involved using radium C and was an objective one, but owing to the technical complexity of the apparatus and agent it is unsuitable for clinical work. Most of the methods described subsequently for the arm–tongue time have been subjective, except those using fluorescein, histamine, and sodium cyanide. In 1933, sodium dehydrocholate was used in a series reported by Tarr, Oppenheimer, and Sager, although its first use is credited to Nobauer in 1923. Saccharin was introduced by Fishberg, Hitzig, and King in 1933. With the exception of Blumgart's radium method, measurements of the arm–lung time depend upon a subjective reaction, and ether, introduced by Hitzig in 1935 and paraldehyde first used by Caudel in 1938, were the agents commonly used. An excellent review of the methods devised was published by Bauer and Slipakoff in 1938.

References to circulation times in congenital heart disease are scanty, but Tarr, Oppenheimer, and Sager (1933), report that the arm–tongue time, in a patient diagnosed as having Fallot's tetralogy, was on three occasions considerably shorter than normal. In 1937, Goldman and McGuire reported the apparent acceleration of blood velocity as measured by the arm–tongue time in three cases of morbus cœruleus. It has been used widely at the Johns Hopkins Hospital, and Taussig (1947) states that the absence of any shortening of the time may support the diagnosis of pure pulmonary stenosis; that shortening of the time may support the diagnosis of Eisenmenger's complex even when there is no obvious cyanosis; and that a prolonged time may be found with aortic stenosis. Prinzmetal in 1941, proposed a quantitative method for the estimates of the actual amount of right to left shunt.

Method Used

The agents used have been 20 per cent sodium dehydrocholate in 18 estimations of the arm–tongue time, and 50 per cent saccharin in a further 22 estimations. Saccharin has been used owing to the recent difficulty in obtaining decholin, although this latter gives a sharper and more distinctive end point and has a lower threshold concentration for taste. In all 35 estimations of the arm–lung time, 5 per cent paraldehyde in saline has been used. The normal range for the arm–tongue time, using these agents, is 11 to 17 seconds, and for the arm–lung time 3 to 8 seconds.

Reports have appeared from time to time of unpleasant effects from the use of decholin, and in three cases these have resulted in the patients' death, though in all the reported cases of death, the patients showed a previous history of sensitivity, such as asthma, etc.; if patients with such a history are excluded from injections, it seems that decholin is a safe substance to use. Saccharin and paraldehyde may cause pain on injection, and the former has been reported as producing abscess formation when injected paravenously. Venous thrombosis has been reported as a frequent complication following saccharin and paraldehyde, whilst nausea and vomiting are said to follow the injection of decholin

on occasions. In this series there has been no fatality, and no evidence of venous thrombosis or any other complication except that about half the subjects complained of pain in the arm following paraldehyde, whilst one vomited after receiving decholin, and another became extremely nauseated.

All patients have been investigated while lying at rest, either in the wards or at the Cardiac Outpatients of Guy's Hospital. The injections have been made with the site of injection, in each case a vein at the bend of the elbow, approximately level with the right auricle. Four ml. of each agent are used, in all-glass syringes fitted with a wide bore intravenous needle. After venepuncture has been performed, a minute or so is allowed to pass whilst the local circulatory conditions return to normal in the previously congested area. This opportunity is taken of ensuring that the patient knows what to expect, and that he has to signal as he tastes the appropriate substance. The time taken for the actual injection has been found to be remarkably constant at 2 to 2·5 seconds, and this can be disregarded in the results. An assistant is instructed to measure the time taken from the beginning of the injection until the patient's signal. Immediately following this, the arm–lung time is estimated, using the same needle, and merely attaching the second syringe. Following the procedure, the patient is instructed to exercise his hand and arm for a few minutes to disperse any paraldehyde that might still be lying in the vein, and so cause thrombosis.

This method is used for the diagnosis of whether a right to left shunt is present or not, but in five cases Prinzmetal's suggested method has been used to assess the percentage of blood shunted from right to left. The method consists, briefly, of the measurement of the arm–tongue time using successively larger amounts of agent. It is only applicable in cases where the shunt does not exceed 50 per cent. The theory upon which the method is based postulates that any substance arriving in the right ventricle (for example) will proceed along the two alternative pathways tongue and lungs (assuming a shunt to be present), in amounts proportional to the volume of blood passing to those organs. There is also a threshold concentration necessary at the taste buds before the subject can appreciate the taste. It follows, therefore, that if less than 50 per cent of blood is shunted into the aorta, the injection of increasing amounts of agent will cause a threshold concentration to be reached first in the blood passing via the lungs to the tongue, and hence the first sensation will be noticed in a relatively long, or normal arm–tongue time. Subsequently, a concentration will be reached in the shunted blood that will produce a taste and when this occurs the arm–tongue time measured will suddenly become shortened.

If the amount necessary to produce the initial longer time be called A, and that required to cause the sudden change, B, then it can be shown that:—

$$\text{Percentage of blood shunted} = \frac{A}{A+B} \times 100.$$

In practice the technique is similar in all respects to that previously described, except that amounts increasing by 0·2 ml. are injected until first the longer, and then the shorter time is obtained.

Results

Of the 36 patients investigated, 35 were suffering from congenital heart disease. The remaining patient (always grossly cyanosed, and for 29 years thought to be a case of Fallot's tetralogy) suffered from pulmonary hæmangioma.

There were 3 patients with congenital heart disease who were not cyanosed.

The average age of patients was 15 years and the range was from 6 to 41 years.

Group I. Cyanosed Patients in whom both Circulation Times were Estimated

There were 28 patients; 20 were shown to have a right to left shunt, and in 8 the times were against a shunt. Details of these 8 are shown in Table I.

The results show failure in 3 (Cases O115, H121, and CB14), and a probable failure in a further one (Case H126) where angiocardiography confirmed the presence of a shunt. In the 4 cases at the top of the table the absence of a right to left shunt was confirmed.

Group II. Cyanosed Patients in whom only the Arm–Tongue Time was Performed

There were 5 patients in this group, and all of these were shown to have a right to left shunt.

Group III. Acyanotic Patients in whom both Times were Performed

There were 2 patients in this group and neither was suspected clinically of having a right to left shunt; in one a patent ductus arteriosus has recently been ligated (Case O122) but in the other angiocardiography very unexpectedly suggested a right to left shunt (Case C206).

Group IV. Patients in whom Prinzmetal's Method was used. (*See Table II*).

TABLE I

PATIENTS NOT DIAGNOSED AS HAVING A RIGHT TO LEFT SHUNT

Reference No.	Sex and age	Arm–tongue (seconds)	Arm–lung (seconds)	Diagnosis	Autopsy
(Baker 1949)	M 29	Decholin 14·0	8·5	Lung hæmangioma	Confirmed No shunt
H117	F 30	Saccharin 17·5	13·0	Valvular pulmonary stenosis	Confirmed No shunt
P215*	F 20	Saccharin 33·0	22·6	Dilated pulmonary artery + aortic regurgitation	Alive No shunt‡
P212†	F 9	Saccharin 16·7	11·0	Valvular pulmonary stenosis	Confirmed No shunt
H126	F 9	Saccharin 13·0	6·0	? Fallot's tetralogy	Alive Shunt present‡
O115	M 27	Decholin 26·0	15·0	Fallot's tetralogy	Confirmed Shunt present
H121	F 24	Saccharin 17·2	9·2	Fallot's tetralogy	Confirmed Shunt present
CB14	F 13	Saccharin 13·8	8·4	Fallot's tetralogy	Confirmed Shunt present

* Cyanosis of peripheral type. † No cyanosis at rest. ‡ Confirmed by angiocardiography.

The result was specially useful in Case P225 as he was not obviously cyanosed at rest; the final diagnosis was Eisenmenger's complex. It was rather surprising in Case 0111 as she too was hardly cyanosed at rest and was thought to have valvular pulmonary stenosis with a patent foramen ovale.

Cases 0070 and 0075 were thought to have Fallot's tetralogy.

Case 0207 was thought to have valvular pulmonary stenosis and to have developed a right to left shunt later in life. This was confirmed at operation and the valve was divided by Mr. R. C. Brock.

She made an uneventful recovery, with striking improvement, already being able to walk about the hospital freely, whereas before she had been virtually bedridden. It has been possible therefore to compare her circulation times before and after operation.

	Arm–Tongue (seconds)	Arm–Lung (seconds)	Hæmoglobin%
Pre-operative ..	12·2	10·0	122
Post-operative ..	10·2	6·8	106

The first difference is the overall reduction in circulation times following operation, coupled with an increase in difference between the arm–tongue and arm–lung times. The lowering of hæmoglobin which occurred may have helped to shorten the times, but it also seems likely that the partial or complete relief of the valvular obstruction has contributed to this effect, and the reduction in pressure in the right ventricle has diminished the shunt which had been taking place through the interauricular septum.

TABLE II

Reference No.	Sex and age	Amount producing longer time	Amount producing shorter time	Shunt
0111*	F 8	variable	results	probably 50%
0075	M 6	0·50 ml.	1·50 ml.	25%
0070	M 8	1·00 ml.	3·00 ml.	25%
0207	F 26	1·80 ml.	3·00 ml.	37%
P225 *	M 13	1·00 ml.	5·00 ml.	17%

* Cyanosis on exertion only.

DISCUSSION

The results presented above have attempted to show that the estimation of the arm–tongue time together with the arm–lung time is a simple and safe method to be used in the diagnosis of the presence of a right to left shunt. It is most important that both times be estimated together, and the only satisfactory evidence of a shunt is that they should agree within two seconds of each other. This alone can indicate that substances introduced into the right side of the heart reach a point on the great and lesser circuits simultaneously, and there must be a communication between the two before the lung capillaries are reached. Further, the blood must be passing from right to left. It is important to note that by no means every case of Fallot's

tetralogy or allied condition shows a markedly shortened arm–tongue time, as might be thought, and indeed as the scanty references in previous work report. In this series five patients had normal arm–tongue times, though in each case the arm–lung time equalled this, which demonstrates the possible error that may occur if the arm–tongue time is taken alone. The chief reason for the failure of the arm–tongue time to be shortened in every case, is probably that the compensatory polycythæmia causes the opposite effect, namely slowing of the time, as shown by Blumgart in cases of polycythæmia rubra vera. In the five cases mentioned here it is noteworthy that the hæmoglobin concentration was 140 per cent or more in each. It is appropriate to comment upon the disparity in actual values obtained for circulation times, using the methods described above and angiocardiography. The normal values stated previously will only hold good if relatively small amounts of agent are introduced into the circulation. Experience with angiocardiograms performed on some of the patients used in this series shows that the opaque dye may appear in the aorta two seconds after injection, giving a grossly shortened arm–tongue time. This is to be explained by the greater quantity of dye injected, namely 50 ml. or more, which consequently causes considerable changes in venous return to the heart.

The method has certain disadvantages. It is, firstly, a subjective method and the patient's intelligent co-operation is needed. This requirement effectively rules out its use in very young children, six years being probably the youngest age at which a satisfactory result may be obtained. Since the patients attending a congenital heart clinic for advice will show a high proportion of young children, many will not be able to undergo the test. Lack of intelligence, even in older persons, apprehension, and general dislike of venepuncture are all factors that have tended to obscure results, and in this series there have been twelve failures from these particular causes.

The results show three subjects in whom a right to left shunt was not suspected upon the circulation times, but in whom autopsy had since shown this contention to be incorrect. These failures all occurred within a short time of each other, when differing amounts of saccharin to be injected were being tested, and the reason for failure seems to be that too little was used. If, as seems reasonable, these patients had a shunt of less than 50 per cent of total, then the injection of too small an amount of saccharin might cause a taste threshold to be reached only by the longer route. There is some support for this, since in the only one of the three cases that had undergone cardiac catheterization, this investigation showed evidence of a moderate shunt only. Since these unsatisfactory results, the amount of saccharin injected has been raised, and no further trouble has been encountered.

Brief experience with Prinzmetal's method has shown this to be tolerably satisfactory, but a rather higher degree of intelligence and co-operation is required. The possible errors are great, and it has been found that repetition of the injection of similar amounts does not always produce the same response. It is unlikely that accuracy to within less than 10 per cent may be achieved, and in any case, in this context, the method is subject to the criticism that providing the presence of a shunt has been diagnosed, the clinical condition of the patient, reinforced by arterial oxygen estimations is of greater importance in indicating urgency of treatment than a doubtfully accurate estimation of the percentage of blood shunted.

Summary

Results of estimating the arm–tongue and arm–lung times in a series of 36 patients suffering from suspected congenital heart disease are given. The agents used were 20 per cent sodium dehydrocholate and 50 per cent saccharin for the arm–tongue time, and 5 per cent paraldehyde for the arm–lung time.

Reference is made to the complications reported by previous workers, and in this series no reactions of any severity were observed. Prinzmetal's method of estimating the percentage of blood shunted in patients known to possess a right to left shunt was used in five cases, and a description of the method is given.

The results show that the measurement of the arm–tongue and arm–lung times together is a reliable, simple, and safe method to be used for the diagnosis of a right to left shunt, and failure was encountered in only three cases. The reasons for these failures are discussed. It is concluded that the single estimation of the arm–tongue time is not a reliable guide to the presence or absence of a shunt. The results using Prinzmetal's method show this to be of doubtful value. A discussion upon the advantages and disadvantages of the method is given.

I should like to thank Dr. Maurice Campbell and Mr. R. C. Brock for their encouragement and advice, and for permission to use their patients for this investigation.

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MORBUS CŒRULEUS

A STUDY OF 50 CASES AFTER THE BLALOCK–TAUSSIG OPERATION

BY

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The Blalock-Taussig operation for morbus cœruleus was described in February 1945. The position reached in September 1947 was presented to the International Conference of Physicians at that time and has since been published by Taussig (1948) and Blalock (1948*a*). The steps that led up to this operation and the subsequent developments have been described recently (Campbell, 1948), so will not be referred to further. The early results of the first 18 operations at Guy's Hospital, some by Mr. Blalock and some by one of us (R.C.B.), have been described (Campbell, 1948); the earlier cases had then been followed for six months.

In the present paper we wish to describe the results in the first 50 patients operated on at Guy's Hospital with a systemic-pulmonary anastomosis for cyanotic heart disease, all since September, 1947, by R.C.B.

All the patients were thought to have Fallot's tetralogy or some closely related form of congenital heart disease, except three (Cases 33, 42, and 49); these three had a similar clinical picture, but with left ventricular preponderance in the electrocardiogram, which nearly always indicates tricuspid atresia or stenosis with a non-functioning right ventricle (Brown, 1936). The earlier cases have now been followed for from 12 to 15 months and the most recent for 6 months. We are not including in the figures that follow 6 patients who have been submitted to operation for pulmonary valvulotomy during this same period.

Age incidence. Most of the patients were between 3 and 15 and there were rather more boys than girls (28 to 22). One was two and a half years old, 5 were three, 32 were between four and ten, 7 between eleven and sixteen, and 5 (by chance, all men) were between 19 and 27 years of age.

Blalock has suggested 3 as the minimum age for operation except in emergencies, and 10 as the ideal upper limit. The operative risk is much greater in patients under 3 and there seems more risk that cyanosis will return, perhaps because the anastomosis fails to grow as the child develops. Partly because these two risks are not likely to diminish suddenly at 3 years of age, and partly because with the large numbers waiting it seems less harmful to delay operation from 3 to 5 than from say 10 to 12, we have tended, latterly, to make 5 years the earliest age.

We have, however, no evidence from our cases that operation is more dangerous or less successful at 3 years of age. We do, so far, find it more dangerous in those over 20, but it will be many years before one can avoid the dilemma of operating on older patients with an increased risk, or allowing them to deteriorate and die.

Symptoms and Signs

Disability. The disability of these patients was extreme: many selected for operation were severe cases who were deteriorating and could wait no longer, rather than good operative risks. Twenty-six were put in the most severe grade (IV) which means that they were made dyspnœic by a few steps and could rarely walk more than 25 yards (Campbell, 1948). One of these (Case 43) said he had once walked 100 yards as a great occasion; as he was 13 and attended an ordinary school in his wheeled chair it emphasizes the disability. Another 19 were in the next grade (III) which means very severe limitation as they could not play outside without frequent rests and could only walk 50 to 200 yards. Three of these, aged 19, 25, and 27, had at one time walked 2 or 3 miles but had become much worse during the last few years.

There were 5 who could do something more than this and were placed in grade II, but even they were very incapacitated. Two, aged 19 and 16, had been

able to walk one or two miles slowly but were most dissatisfied with being "unable to do anything" and were enthusiastic for operation, whatever the risks. The other three were younger and could walk half to one mile on a good day, but often they could not do as much as this. As further evidence that even these were moderately severe cases, all except one had hæmoglobin percentages between 150 and 126.

The large proportion of older patients among the last eight suggests that they had survived because their condition was less severe but was now deteriorating. All cases of Fallot's tetralogy have not, of course, such severe disability, but the worst have been chosen for early operation.

In addition, some of these patients were becoming rapidly worse so that operation was expedited. For example, Case 46 attended a second time after three months: the distance she could walk had shortened from 150 to 50 yards; her cyanosis and the clubbing of her fingers had become worse. The slightest exertion, even dressing, provoked almost daily attacks of loss of control of her limbs with semi-consciousness.

Our experience suggests that when a patient with Fallot's tetralogy starts deteriorating the prognosis is grave, and several times when this has happened death has not been long delayed.

Cyanosis. All these patients had been cyanosed from early infancy and all had clubbed fingers. The measurement of cyanosis is difficult and is made more so by its quick variation with exertion and temperature. In bed in a warm ward many of the patients look so much less blue than as out-patients that they are hardly recognizable. With Dr. W. D. Brinton we made some attempts at colour matching but they have not so far been successful. The relationship of cyanosis and polycythæmia is reciprocal, the lack of oxygen causing the polycythæmia and this in its turn increasing the appearance of cyanosis.

The estimate of cyanosis was made without knowing the hæmoglobin percentage. In general terms, those in whom it was from 110 to 129 had been placed in grade II or III as regards their cyanosis; those from 130 to 139, in grade III; and those from 140 to 160, in grade III or IV. These and some other details are given in Table I. But there was no very close correspondence and sometimes rather surprising contrasts.

The cyanosis was generally severe (grade III,* 25 cases) or very severe (grade IV, 15 cases). In 9 it was less than this and often less than would have been expected from the disability, though it was always present even with the patient at rest: in these 9 the hæmoglobin averaged 122 per cent.

In one patient in particular (Case 26) where the disability was very great, the cyanosis and clubbing slight, and the hæmoglobin 116 per cent, we were somewhat hesitant about operation but the result was just as successful as in others. The arterial oxygen saturation was 80 per cent falling to 75 per cent with very trivial leg movements (Dr. Zak). If the disability is severe enough, relatively slight cyanosis should not as a rule be a contra-indication to operation.

Onset of cyanosis. The age from which cyanosis was first noticed is of great importance. Naturally, if there is an over-riding aorta and a ventricular septal defect, venous blood will be passing into the aorta from birth or soon after. Cyanosis may be noted at once depending on the amount of shunt, and will become more obvious as greater demands are made on the circulation and as polycythæmia develops. It is most important not to mistake temporary cyanosis *at birth* for cyanosis that has persisted *from birth.*

Cyanosis was noted from an earlier age than 18 months in all except one. The actual figures were from birth or soon after in 30 of the 50, from between 2 and 6 months in 9, from between 7 and 10 months in 6, from between 15 and 18 months in 4, and at 24 months in Case 32.

Of the 5 where cyanosis was not noted till after 15 months, 4 seemed ordinary cases of Fallot's tetralogy though one lived to 27: the fifth (Case 32) had pulmonary stenosis and transposition of the aorta so that cyanosis might have been expected from birth. It is strange, but can hardly be more than a coincidence, that 3 of these 5 patients died after operation.

Polycythæmia. The hæmoglobin percentage varied from 110 to 160 and averaged 137. The highest figures were 160 (Case 35), 158 (Case 29), 157 (Case 6), and 153 (Case 22). All these except the first looked severely polycythæmic.

The red blood cells were generally between 6·0 and 9·0 and averaged 7·8 millions, but in two it was 11·6 and 11·1 millions (Cases 41 and 47), the next highest being 10·1 million and several of 9·0 million or just over. Curiously enough these two had not specially high hæmoglobin percentages, the figures being 140 and 137 so that the colour indices were unusually low—0·58 and 0·60 respectively.

The hæmatocrit reading was generally between 60 and 85 and averaged 74. As might be expected, it generally agreed more closely but not very closely with the red cell count. The highest readings were 94 (Case 41, one of the highest red cell counts), 92 (Case 29, one of the highest hæmoglobins), 88, 87,

* Grade III. Cyanosis moderately severe *at rest* and obvious at a glance. Grade IV. Cyanosis gross, *at rest.*

TABLE I

CASES OF MORBUS CŒRULEUS SUBMITTED TO BLALOCK-TAUSSIG OPERATION AT GUY'S HOSPITAL

Case No.	Initials	Sex and Age	Cyanosis	Disability	Hæmoglobin (percentage)	Red cells (millions)	Hæmotocrit	Hæmoglobin 3 or 4 weeks after operation	Reference No.
1A	A.C.	M 19	4	3	148	10·0	78	Died	P01A
19	S.F.	M 4	2–3	3	112	6·0	58	100	CB04
20	M.E.	F 5	2–3	3	110	5·9	63	100	0018
21	T.G.	M 4	3	3–	138	8·0	(68)	127	C004
22	M.L.	F 13	4	4	153	7·2	87	N.C.*	C005
23	D.S.	M 19	4	4–	130	6·0	77	106	0015
24	C.M.	F 6	4	4	148	8·8	81	128	0019
25	J.R.	F 6	2–3	4	140	7·4	70	101	0005
26	J.L.	M 15	2	4	116	8·4	57	88	P069
27	D.F.	F 10	3+	3	150	8·0	83	N.C.	0027
28	J.L.	F 3	4	4	107	6·6	64	N.C.	H113
29	T.H.	M 10	4	4	158	8·2	92	122	0036
30	G.F.	F 7	3–	3–	133	8·0	72	—	P045
31	V.S.	F 2	2–3	2–3	136	7·4	72	110	C008
32	J.H.	M 11	3–	4	144	8·0	74	Died	P054
33	P.S.†	M 4	3	4	132	9·2	85	109	0022
34	A.W.	F 9	3	2+	118	6·4	63	97	H128
35	M.C.	F 7	3	4	160	7·9	67	Died	P037
36	P.R.	F 16	2+	2+	126	5·3	58	89	C007
37	J.W.	M 5	4	4	136	8·4	81	103	P102
38	C.B.	F 7	3–	4	142	10·1	81	Died	H106
39	P.C.	M 10	3	3	139	8·3	87	114	CB09
40	D.S.	M 5	3+	3	140	7·7	71	105	P089
41	J.H.	F 5	4	4	140	11·6	94	115	CB17
42	S.H.†	M 19	3	2+	144	8·0	—	112	P057
43	T.H.	M 13	3	4	142	7·2	69	—	P056
44	G.S.	F 11	4	4	142	8·6	84	110	H120
45	S.N.	M 3	2	4	109	6·6	—	92	0017
46	K.C.	F 15	3	4	135	8·9	76	—	0105
47	G.K.	M 11	3	3	137	11·1	—	115	0184
48	R.B.	M 27	4	3	144	9·1	85	Died	0115
49	J.C.†	M 8	4	4	125	8·6	73	123	0007
50	M.S.	F 13	3	2+	138	7·2	—	110	0140

* N.C.=No change as no effective anastomosis. † Tricuspid atresia.

Cases 1–17 have been described previously (Campbell, 1948).

and 86; several others were very close to this.

As would be expected, the red cell count increases regularly with the hæmoglobin percentage in the lower ranges, but to our surprise this parallel increase is not continued in the higher ranges. In Table II the cases have been classified according to the hæmoglobin and it will be seen that the red cells and hæmatocrit increase as the hæmoglobin goes up from 112 to 134, but there is no further increase either in the red cells or in the hæmatocrit as the hæmoglobin goes up from 134 to 153. The disadvantages of a red cell count over 8·0 million may produce some mechanism in the body that prevents a further rise, but if so the two cases with counts over 11·0 million are all the more surprising.

Apparently as the hæmoglobin rises above 130 per cent the red cell count and hæmatocrit do not on the average rise further with the result that the colour index rises towards unity.

Clubbing of the fingers and toes. All these cases showed moderate or severe clubbing of the fingers and toes, and Case 26 (whose hæmoglobin was 116 per cent) was the only one who did not have the complete picture as he had curvature of the nails without any noticeable broadening. Fifteen were marked as having moderate rather than severe clubbing and these included 8 with hæmoglobin percentages of from 110 to 126, but the others were about 140. Six were marked as having unusually severe clubbing: their hæmoglobin ranged from

TABLE II

AVERAGE BLOOD COUNTS GROUPED BY THE HÆMOGLOBIN LEVEL

Hæmoglobin		No. of cases	Average red cell count (millions)	Average hæmato-crit	Average colour index
Range	Average				
110–119	112	8	6·5	62	0·86
120–129	124	4	7·2	63	0·86
130–139	134	12	8·3*	77	0·81*
140–149	145	16	8·2	80	0·88
150–160	153	10	7·9	76	0·97
110–129	116	12	6·7	63	0·86
130–160	144	38	8·1	78	0·88
All cases	137	50	7·8	74	0·87

* Without the two cases with counts over 11·0 million these figures would be 7·8 and 0·85.

130 to 160 per cent. This correlation between clubbing and polycythæmia might be expected. Fig. 1 and 2 show severe clubbing of the same fingers a year after operation when the degree of clubbing has become much less.

Squatting. Of these 50 patients, 41 had a history of squatting and 7 gave no such history; there were 2 with no information. Fig. 3 illustrates the typical position. Taussig considers squatting almost constant in Fallot's tetralogy, but our evidence is against this though we find its does occur in about 80 per cent. It seems unlikely that the diagnosis was wrong in 7 who were not squatting. One had tricuspid atresia (Case 42) and the operation was as successful in him as it was in 3 others who did not squat (Cases 5, 10, and 36). It is curious, but again perhaps no more than a coincidence, that 3 of the relatively small number of unsuccessful results (Cases 11, 22, and 27) occurred among this small number of non-squatters.

Mental and physical development. It is surprising that the mental development should be so good, considering the severity of the anoxæmia. All but two of these children were normal or often above the average, although, of course, many were educationally backward. We have used the age of walking and talking as rough measurements.

Generally the child walked at a normal age but a few were late. Most children (33 cases) started walking at or before two years, but it was delayed in 12 till three years, in 2 till four years, in 1 till five years, and in 2 till seven years. There was no mental defect in these children and the last two boys seemed of average or more than average intelligence.

Most children (38 cases) started talking at a

FIG. 1.—Clubbing of fingers and its disappearance after operation. This position shows the disappearance of the broadening from side to side and some diminution of the curvature. With moderate clubbing it may disappear completely in five or six months but with a more severe grade such as this, something is left permanently and the illustration is nearly a year after operation. Case 23.

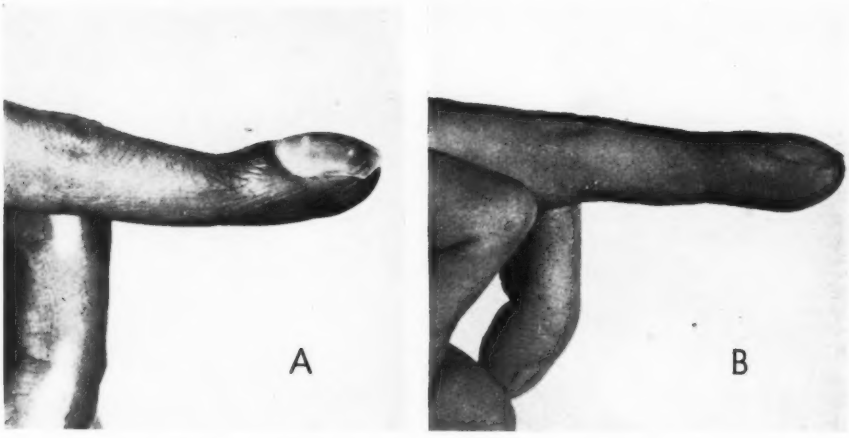


FIG. 2.—Clubbing of the fingers before and a year after operation, showing diminution of the broadening from front to back and of the curvature, and a more normal appearance of the skin. Case 23.

normal age, generally between 12 and 18 months. Seven were said to have been able to talk at two years, 2 at two and a half years, and 3 at three years.

Only 4 (Cases 19, 28, 35, and 49) were slow both in walking and talking. Case 19 was mentally backward and Case 35, who had a cataract also, following maternal rubella during pregnancy, was rather backward. All the others were normal mentally.

Most of the children were under weight and their parents always complained of difficulty in getting them to eat. On the average they were only slightly under height so that their thinness was very obvious. Often the chest was badly developed with some tendency to pigeon chest and to a Harrison's sulcus. The veins over the chest were often more visible than normally.

PHYSICAL SIGNS

The heart is generally of normal size and anything more than very slight enlargement should raise a suspicion that the lesion is not Fallot's tetralogy or is complicated by the addition of some other defect. The size and shape is dealt with more fully in the section on radiology.

Systolic murmur and thrill. There is generally a *systolic murmur* in the second, third, and fourth spaces on the left, becoming fainter towards the apex. The murmur is often loud enough to be conducted widely, sometimes to the right side or to the back. Sometimes the pitch of the murmur differs over the pulmonary area and lower down in the fourth left space and this may be due to there being two different murmurs produced by the pulmonary stenosis and by the ventricular septal defect.

The loudness and harshness of the murmur varies a great deal. There were 4 cases where no systolic murmur was heard (Cases 9, 22, 41, and 48); all were of severe degree and in the last, who died, there was a calcified pulmonary valve. In 2 of these 4 and in 2 others, triple rhythm with an addition of a third heart sound was so obvious as to be almost the main physical sign. It was equally common for the murmur to be described as soft or faint (13 cases); as average (13 cases); or as loud, rough, or harsh (14 cases + 6 special cases). The 6 special cases with a harsh or rough murmur were the 3 with tricuspid atresia, Case 28 with a large pulmonary artery, and Cases 32 and 35 with unusual features discovered post-mortem (see page 192).

Such a harsh or rough murmur (or thrill of well-marked intensity) is, therefore, a reason for considering carefully if the diagnosis is correct, though it may be found fairly often in an ordinary case of Fallot's tetralogy, perhaps indicating that the stenosis is fairly severe; but not that there is atresia, when a murmur may be absent.

In none of these cases was any diastolic murmur heard, but some others with the features of Fallot's tetralogy and a diastolic murmur have been deferred for fuller investigation.

A *thrill* was present in more than half, but generally it was faint and might only be felt at times or after exertion. It was usually maximal

FIG. 3.—Case 33 with tricuspid atresia and non-functioning right ventricle. A typical position adopted in squatting, though in many of the older patients the knees are brought still closer to the chest.

in the pulmonary area but occasionally towards the apex. As would be expected it was closely correlated with the harshness of the systolic murmur. The thrill was never of great intensity; it was of moderate intensity in 13 cases, but this included the with tricuspid atresia and the 2 with unusual findings post-mortem. It was difficult to feel or only felt occasionally or after exercise in 17 cases. No thrill was felt at the many examinations in 0 cases.

Pulmonary second sound. Auscultation should be carried out with the patient sitting up and lying down and during each phase of respiration, before a decision is taken about the intensity of the second sound.

Diminution of the second sound in the pulmonary area has been traditionally regarded as an important sign in the diagnosis of pulmonary stenosis. As far as Fallot's tetralogy is concerned this is not so; the second sound is generally normal and is as likely

to be a little increased as diminished. It was often as loud on the left as on the right side at the base.

Any great increase, however, is likely to indicate that the pulmonary pressure is raised and that on screening a large pulsating pulmonary artery will be seen and that the lung fields will be congested instead of clear. This is specially true if the second sound has a drum-like quality. Visible pulsation in the pulmonary area, and palpable diastolic shock also suggest that the lesion is not Fallot's tetralogy and that the pulmonary pressure is raised. We would emphasize that the significance of the change in the pulmonary second sound depends on a sound that is *much increased* and not to one that is *slightly increased*.

In these 50 cases it was recorded as normal in 17, as diminished in 11, and as slightly increased in 13; in 9 it was more notably increased. These 9 included one where the pulmonary pressure was high (Case 28), one with tricuspid atresia (Case 42), one with severe cyanosis who was hardly helped by operation (Case 22), and one who had infundibular stenosis and a transposed aorta (Case 32); but the other five seemed ordinary cases who were helped by operation.

No case of Fallot's tetralogy has had a drum-like pulmonary second sound, even if the pulmonary artery was more prominent than usual. A diastolic murmur immediately after this sound was never heard in Fallot's tetralogy though both these findings are not uncommon in other types of cyanotic congenital heart disease.

Blood pressure. The blood pressure in these cases averaged 106/73, though sometimes it was hard to get an accurate diastolic reading. In nearly every case it was within the range 115–95/80–65 and in 4 cases where it was about 127/90, all were over 15 years of age. In some of the older patients it seems to be increasing a little, still with a small pulse pressure.

Radiology of the Heart

In the account of the first 18 cases (Campbell, 1948), the size and shape of the heart was discussed at some length and the difficulty of describing any characteristic shape was emphasized. Less than half had hearts that were sabot shaped and the other half had more normal shaped hearts; sometimes with a gross hollow pulmonary bay, but often with an almost straight left border or occasionally even with some slight prominence in the region of the pulmonary conus. The findings are much the same in the present series and we are not discussing the question further.

We would, however, emphasize three negative points as of the greatest importance.

The density of the lung. The most decisive—and perhaps the most decisive point in the diagnosis of a condition that can be helped by systemic-pulmonary anastomosis—is the absence of noticeable pulsation in the lung roots, with an absence of density in the lung fields as a whole.

The size of the heart. The second important point is the size of the heart. In most cases of Fallot's tetralogy, however great the disability and cyanosis, the heart is of normal size or even smaller. Anything more than trivial enlargement of the heart makes one hesitant about operation, partly because it suggests that there are greater complications in the congenital abnormality, and partly because the heart is less able to stand any enlargement that may follow the creation of an artificial ductus arteriosus. This is discussed more fully in the next section.

The pulmonary artery. The third point, of almost equal importance, is that there should be no undue prominence of the pulmonary artery and better still, that there should be a striking hollow in the pulmonary region, though as already stated, some patients have a rather straight left border. There may even be a convex projection in the region of the conus just below the origin of the pulmonary artery due to the prominence of the infundibulum distal to the infundibular stenosis. Or there may be a dilatation of the pulmonary artery beyond a pulmonary valvular stenosis but this is rare with Fallot's tetralogy, and should show no pulsation.

It is desirable that one should be able to see both pulmonary branches, because then a pulmonary vessel is available for the anastomosis and there is no risk of the patient dying suddenly from arrest of the pulmonary blood flow when one pulmonary artery is clamped. This accident occurred in Case 17; in Case 27 no operation was possible because the pulmonary artery was too small. Looking back at the X-ray films we think that in the latter, it should have been possible to tell this before operation, but in the former it would not have been easy, as there was relative density round the lung roots, presumably owing to the collateral circulation.

In many cases, the hollow in the region of the pulmonary artery in the P–A view and the large aortic window in the left oblique, together with the absence of pulsation and the absence of density in the lung fields, make the diagnosis easy after radioscopy, and there seems no need for any further investigation. In others the prominence of the pulmonary artery and the density of the lungs, with or without pulsation far out in the lung fields, at once makes it obvious that the patient is not suitable for operation. But there remain others where the decision is difficult; the density produced by the

collateral circulation causes one of the greatest difficulties but here there is no pulsation and more pin-point scattered shadows. Our first mistake in this direction was in Case 28 where we had been doubtful about the prominence of the lung fields but had decided there was pulmonary stenosis. However, at operation the pulmonary artery was found to be large and pulsating with a pressure well over 80 mm. of mercury. This was in a child who was unable to stand and had been screened lying down.

The right (I) oblique position on radioscopy will help in showing if there is any undue prominence or pulsation of the pulmonary artery or of its left branch. The left (II) oblique will define the relative size of the ventricles and will generally show the left of normal size with the right somewhat, but generally not greatly, enlarged. It also helps to show the size of the pulmonary artery and of the aorta.

The aortic arch. A barium swallow is necessary to determine whether the aortic arch is on the left or right. As the barium is swallowed it is often of help to keep ones eyes fixed on the aortic knuckle, as the barium may sometimes follow a preliminary curve to the right before it reaches the aortic knuckle and if this is small it may be missed. It is hardly necessary to say that the barium should be of thick consistency.

The aortic arch was right-sided in 14 and left sided in 36 of the cases which is nearly the usual proportion of 1 in 4.

The Size of the Heart

The heart size is not easy to estimate with accuracy, and general opinion ranks inspection of the film and still better, of the heart on radioscopy, as a better method than any specific measurement.

The following estimates of the heart sizes have been decided by one of us (M.C.) in retrospect, mainly on the P–A films, because these are easiest for comparison from case to case and were always available. Some attention was paid to the cardio-thoracic ratio (c.t.r.) in each but the decision was made mainly on the general appearance. It is, therefore, interesting to see how these estimates compare with the c.t.r. which are more useful for conveying an idea to others. They are given below against the estimated size of the heart; the figures in brackets refers to the number of cases.

Very small	37–43 (4)
Small	41–49 (10)
Small normal	45–50 (5)
Normal	45–52 (15)
Large normal	49–54 (8)
Enlarged	52–61 (8)

In 14 cases the hearts were regarded as small and in 8 only as enlarged, though possibly some people might have counted the 8 "large normal" as enlarged: the remaining 28 (or 20) were regarded as of usual size.

The 8 that were enlarged will be discussed in more detail, and taking them from the largest downwards they were as follows:

Case 35 (c.t.r. 62) died after operation and there was a single auricle as well as the other features of Fallot's tetralogy.

Case 33 (c.t.r. 61) had tricuspid atresia with a non-functioning right ventricle and although the operation has been most successful and the patient is able to lead a life quite unlike anything previously, the heart has subsequently enlarged more than we like.

Case 28 (c.t.r. 56) had some doubt about the lung fields before operation, and at operation the pulmonary artery was found to be large with a high pressure and the patient probably had Eisenmenger's complex.

Case 11 (c.t.r. 55) was not helped by operation but we thought this was due to technical difficulties with the anastomosis and did not indicate a wrong diagnosis.

The next four fall into a category of slighter enlargement.

Cases 8 *and* 47 (c.t.r. 52 and 53) seemed to be ordinary cases of Fallot's tetralogy and the operation was satisfactory, Case 8 being 25 years old.

Case 48 (c.t.r. 52) was an ordinary case of Fallot's tetralogy who died after operation; he was 27.

Case 32 (c.t.r. 52) had the aorta arising from the right ventricle and he, too, died after operation. It will be seen that among these 8 cases there were several where the diagnosis was more complicated than straightforward Fallot's tetralogy or where the patients did not do well, and the greatest care should be exercised in chosing any patient with significant cardiac enlargement.

There were four other patients where we decided to call the heart large normal, though it was difficult to be sure it was not slightly enlarged; it was interesting that the c.t.r. were much the same as the four just described, where we thought there was enlargement.

Cases 45 and 30 (c.t.r. 54 and 53) seemed to be ordinary examples of Fallot's tetralogy and did well after operation. Case 38 (c.t.r. 53) who died after operation had Fallot's tetralogy and Case 49 (c.t.r. 52·5) had tricuspid atresia and did well after operation. As three of these did well and there is no reason to think that the one death was due to the heart size, it is probable that similar cases can reasonably be included as suitable for operation

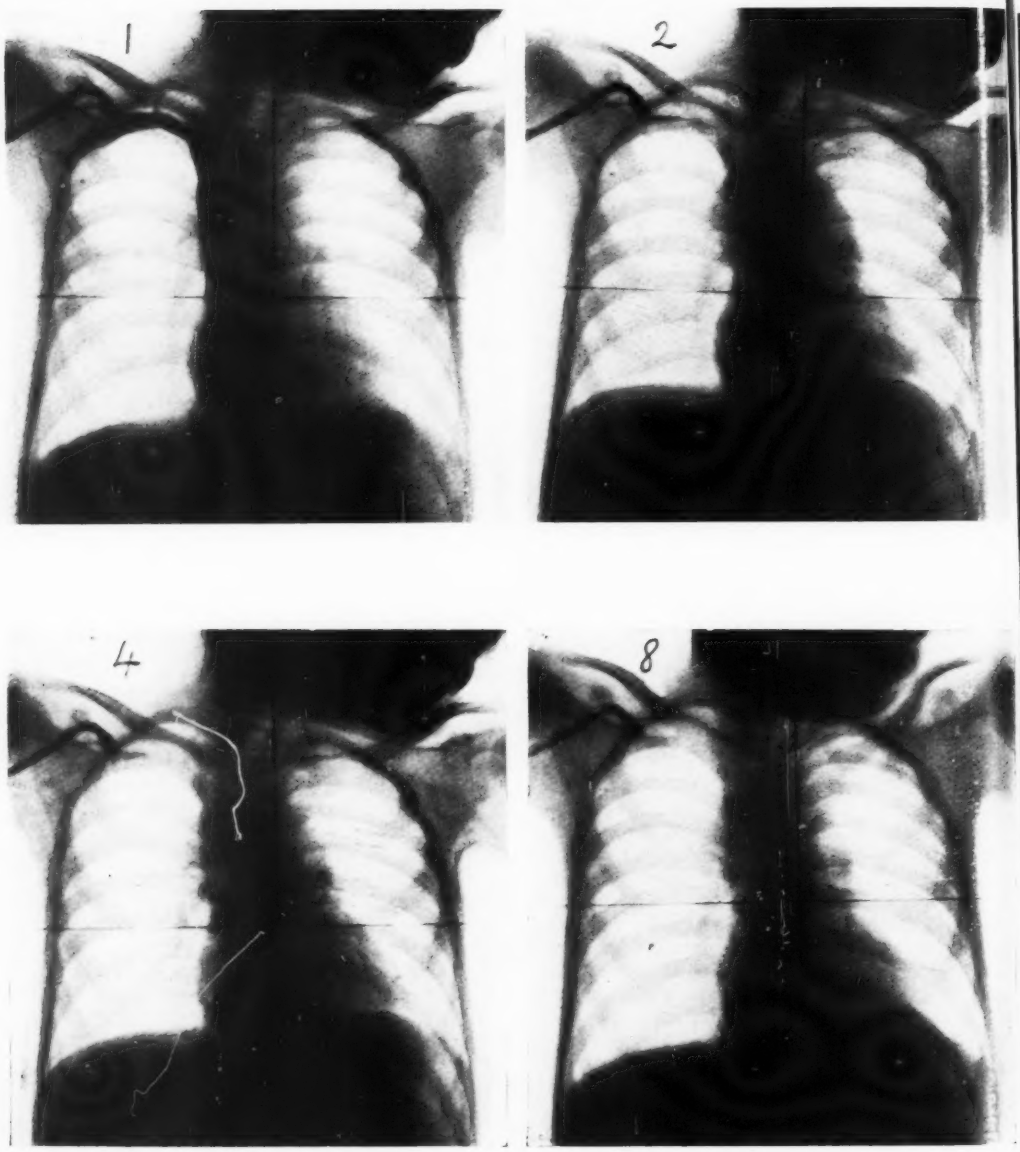


FIG. 4.—Case 39. Fallot's tetralogy. The film at 1 second shows normal filling of the right auricle. The film at 2 seconds shows striking filling of the large aorta arching to the right and some filling of its branches. There is evidence of filling of the left ventricle. There is no significant change in the pulmonary arteries. The film at 4 seconds still shows the aorta but less clearly and the subclavian more clearly. The filling of the pulmonary arteries is trivial but this was the maximum reached. In the film at 8 seconds the shadows are fading though the aorta can still be seen. The pulmonary arteries and the lungs as a whole are lighter than at 4 seconds. The conclusion is a large right to left shunt with a right-sided aortic arch and a fairly severe pulmonary stenosis.

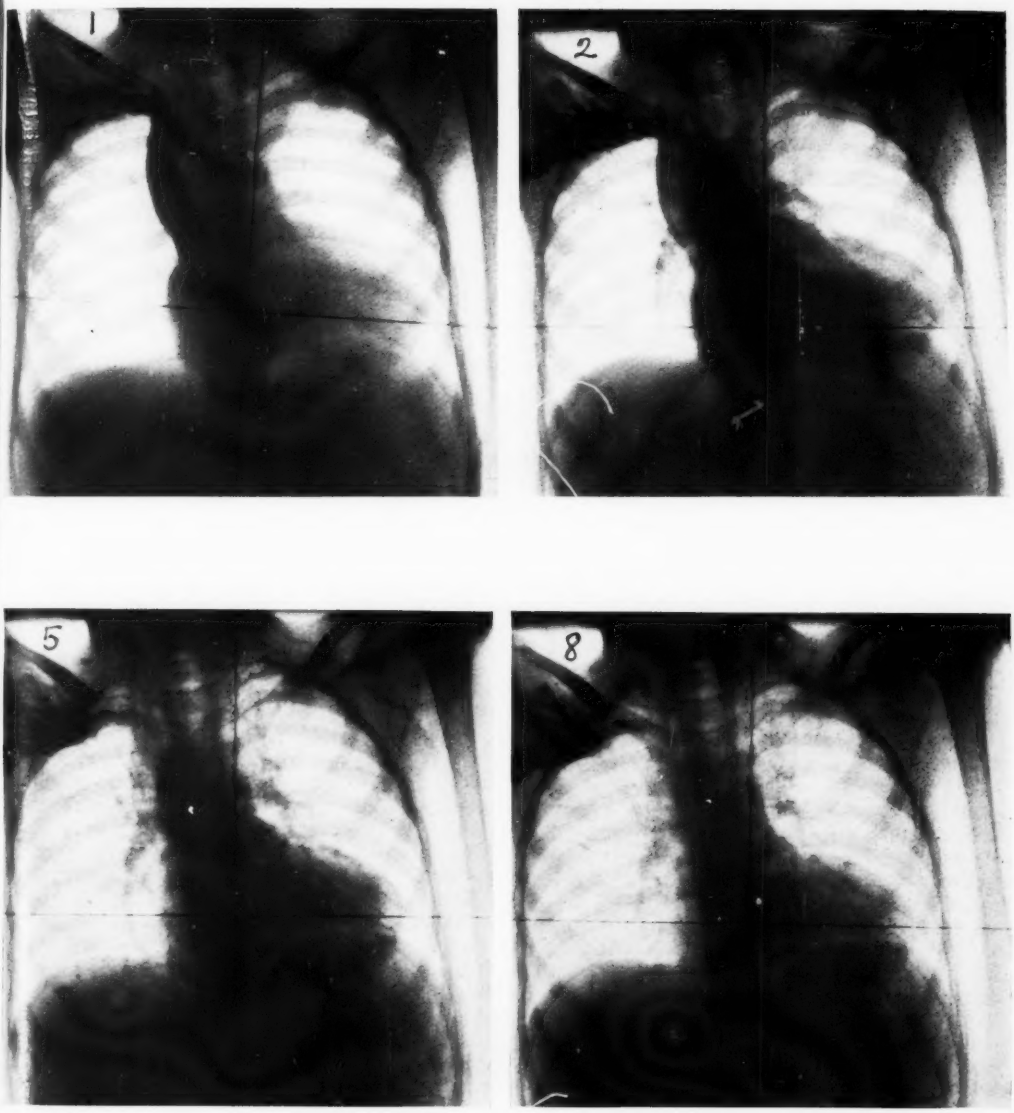


FIG. 5.—Case 47. Fallot's tetralogy. The film at 1 second shows normal filling of the S.V.C. and right auricle. The film at 2 seconds shows dense filling of the right ventricle and of the aorta arching to the right and of its branches as far as the subclavian in the left axilla, indicating an aorta in free communication with the right ventricle; the dye has not passed out to the borders of the left ventricle. At the same time, the pulmonary arteries have begun to fill. The film at 5 seconds has been chosen because it shows as much pulmonary filling as at any time and obviously little blood has entered the lungs. The degree of filling is a far better indication of pulmonary stenosis than the time at which dye can first be seen in the pulmonary arteries. The right ventricle and aorta are clearing. The film at 8 seconds shows that the heart as a whole is becoming clearer. The conclusion is a large right to left shunt with a moderate degree of pulmonary stenosis.

and that if the heart can be passed as not much enlarged a relatively high c.t.r. is not a reason for deciding against operation.

We have no example to report of a successful operation in a patient with Fallot's tetralogy with the c.t.r. over 55. But this may be partly due to selection of patients who had not the larger hearts, and this is a question that still needs more experience. We are glad we did not exclude the boy with tricuspid atresia with c.t.r. of 61 but whether his improvement can last as long as in some of the others remains to be seen.

From our present experience and from the tables of Lincoln and Spillman (1928), of Maresh and Washburn (1938), and of Caffey (1945) it seems that under the age of 2 years the c.t.r. averages 49 and varies between 40 and 65 (60 after one year) in normal children. These figures do not concern our present purpose directly but may help in advising parents whether their children may possibly be helped by operation later.

From 2 to 5 years of age the normal range lies between 43 and 52, and from 6 years onwards the average c.t.r. falls slightly from 47 to 45, with a range from about 40 to 50. The original figures of Danzer as long ago as 1919 gave 39 to 50 as the adult range with an average of 45; he stated that a c.t.r. of 52 might be normal if the heart did not look enlarged, but that one of 53 was pathological. These last figures were of course concerned with adults but seem to agree with our conclusions.

On our present experience, we consider that a heart which seems a little enlarged with the c.t.r. of 52 to 54 should certainly not contraindicate operation, though a larger heart than this often indicates the presence of some complication and is probably a bar to *lasting* improvement. We have not yet sufficient evidence to say how often operation should be advised in these larger hearts for the sake of immediate advantages.

Angiocardiography

We do not propose to discuss in detail the help that can be obtained from angiocardiography which was only available in nine of the later cases of this series. As a rule, there should be no need for this help from the point of view of diagnosis, but in border line cases where a heavy collateral circulation hides the diminished blood supply to the lungs, it may be of the greatest value; and it may help to establish the diagnosis in complicated cases where no diagnosis can be made on clinical grounds alone. Apart from this, it has proved of increasing value from the surgical point of view in delineating the anatomical arrangement of the arterial branches from the aorta and of the size and position of the pulmonary arteries.

As Fig. 4 and 5 show, there is no difficulty in demonstrating the shunt from the over-riding aorta and this is generally well seen in the film taken at the 2nd second in cases of Fallot's tetralogy. Often the pulmonary arteries start filling at the same time, which might suggest there was no great degree of pulmonary stenosis but we have found that the amount of the increased density of the lungs during the subsequent 5 seconds is a better test than the speed with which the opaque substance can first be seen in the pulmonary arteries. Even so, angiocardiography seems to give an added precision to the assessment of the degree of pulmonary stenosis present in different cases of Fallot's tetralogy, and may help in distinguishing between valvular and infundibular stenosis. Both these points may ultimately be useful in deciding the sort of operation that is most likely to be successful.

Electrocardiograms

The two most striking features of the cardiogram are the large pointed P wave especially in lead II and the right ventricular preponderance. This is of such a degree that we think the term ventricular preponderance rather than axis deviation is justified even on the standard leads. We hope to deal later with the value of unipolar chest leads as these were not available for all the early cases.

Wood and Selzer (1939) thought that a tall spiked P wave might be produced by a right auricular hypertrophy. Pardee (1941) accepts the view that abnormally high pointed P waves occur with hypertrophy of the right auricle, while notching and broadening are seen with hypertrophy of the left auricle.

The prominent pointed P wave was generally tallest in lead II and some examples are given in Fig. 6. The width of the P waves was generally 2 mm. and exceptionally up to 3 mm. In these 50 cases P II varied from 2 to 8 mm. in height, generally between 3 to 7 mm. Once (Case 30) it was small and sharply inverted. There was only one other where P II was as small as 2 mm. high, 10 cases where it was 3 mm. high, 16 where it was 4 mm. 11 where it was 5 mm., 6 where it was 6 mm., 4 where it was 7 mm. and 1 where it was 8 mm. high. The average was 4·5 mm. Chamberlain and Hay (1939) give 1·5 mm. as the average size of P II in the first decade with a maximum of 3·0 mm. Pardee (1941) gives 2 to 5 mm. as the usual size for the large P waves of mitral stenosis.

The cause for these large P waves is not certain. It is not due to the tachycardia that is generally

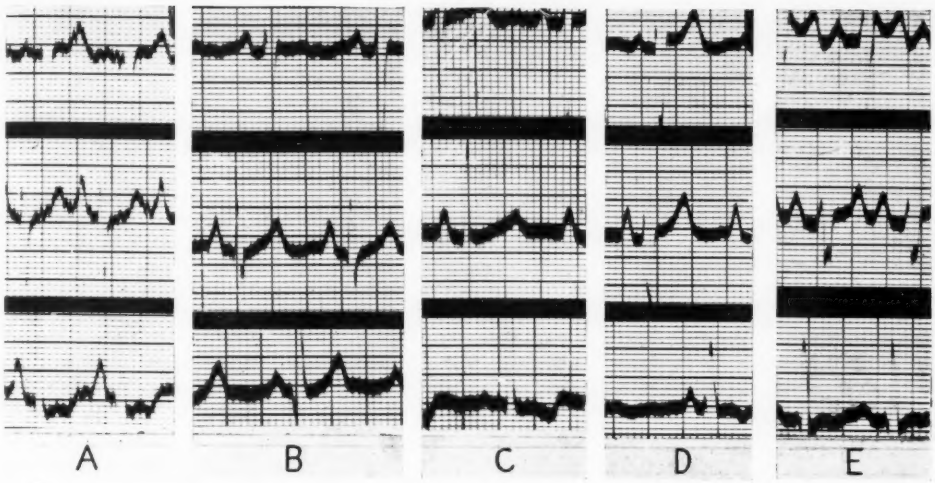


FIG. 6.—Typical electrocardiograms from 5 cases of Fallot's tetralogy showing gross right axis deviation and large pointed P waves. (A) from Case 0208 confirmed post-mortem. (B)–(E) from Cases P067, H120, P022, and P035 confirmed by a successful result of operation for a systemic pulmonary anastomosis. (Reduced to five-sixths.)

present, especially in the younger patients, as equally tall P waves occur in the same subject when the rate is slower. The variability suggests that it is partly due to strain on the right auricle as well as the hypertrophy which should make for greater constancy. Moderately large and pointed P waves may occur with cardiac infarction or in cor pulmonale where there is right heart strain. Large P waves were seen equally in the cases of tricuspid atresia.

In Case 35 where there was a single auricle the P wave was large and pointed, 5 mm. high. In Case 49 with tricuspid atresia it was tall and bifid as well. In the other two cases with tricuspid atresia the P wave was sometimes large and broad in Case 42, but in Case 33 it was not remarkable. Occasionally the P waves may show a fair amount of variation in height in the same record (e.g. Case 35). Where more than one cardiogram was taken before operation the P waves often differed in amplitude but never to any great extent except in Case 36; although there were large pointed P waves in her first cardiogram with a rate of 115, a subsequent one done six months later, but before operation, showed normal small P waves, with a rate of 75; there were no other significant changes in the two curves.

The most significant finding as one might expect was the high degree of right ventricular preponderance. The main exceptions were the 3 cases with left ventricular preponderance; it is fairly certain that such a case will turn out to be tricuspid atresia or stenosis with a non-functioning right ventricle, especially if on radioscopy the left is the larger of the two ventricles.

In the 47 cases that were thought to have Fallot's tetralogy, the general finding was an absent Q, a diminutive or very small R and a large S in lead I; and an absent or very small Q, a large R and most constantly of all, an absence of S in lead III. Out of these 47 there were only 4 with S III and in these it ranged from 3 to 8 mm. in depth. The average figures for the size showed that R I, Q III, and S III were small, 2·8, 1·7, and 0·4 mm. respectively, while S I and R III were large, 12·9 and 13·3 mm. respectively.

The QRS complexes in lead II may be of the R type (23 cases) or S type (15 cases) or mixed (9 cases) or with very small QRS complexes (4 cases).

Now and again one meets with a case that does not show a high degree of right preponderance. Case 6 is an example where there was no preponderance possibly indicating a single ventricle as well as the usual features of Fallot's tetralogy; the result of operation was very successful but the colour was not improved as much as in most cases. In Cases 1, 30, and 37, all successful operations, there was less right preponderance than usual. Some cases showed a moderate R I in addition, of course, to the very deep S I.

T wave abnormalities were uncommon except for T III inversion: in 11 cases it was sharply inverted and in 7 it was flat or slightly inverted. There were no T II inversion except in Case 30 where it was biphasic.

In two, T I was inverted (Cases 42 and 47) and in one it was flat (Case 17). In Case 42 for some unknown reason it became normal and upright again six months later, before his operation. In Case 21, T I was enormous, 11 mm. in amplitude, and T III was deeply inverted. Fairly large T waves in lead II with an amplitude of over 5 mm. occurred in 8 cases.

As with the P waves, the T waves in some of these cases showed variation in height, unlike most other types of heart disease where the size and shape remain remarkably constant. The S–T interval was often elevated in lead I and sometimes became more normal after operation.

Operative Experiences

In these first 50 cases the original basic technique, as recommended by Blalock, has been closely followed; in 6 other cases valvulotomy was performed or attempted. Pott's modification (anastomosis of aorta to pulmonary artery) was not used in this series, although it has been since.

The Blalock operation of anastomosis of a systemic artery to a pulmonary artery is always difficult and exacting technically, even when the anatomical conditions are favourable and the anastomosis proceeds smoothly. At times, when conditions are not favourable, it becomes an operation of really great difficulty and calls for all the surgeon's technical ability. Some of the earlier more difficult cases were especially exacting, and even with increased experience it is found that a series of straight-forward operations is suddenly interrupted by a complex anatomical situation calling for great patience and not a little endurance.

This is not an operation for the casual operator and indeed, quite apart from the pressure of the waiting list, it is desirable that the surgeon should operate regularly. In Blalock's clinic one operation a day is aimed at; it has not been possible to achieve this here because routine thoracic work must still be done, but a desirable standard should be two or three operations a week. Success is not possible without good team work and one must stress in particular the invaluable part played by the anæsthetists; Rink, Helliwell, and Hutton (1948) have already written a preliminary record of their experiences.

Often, these cyanotic patients are not in the state of general health that one would ordinarily demand before embarking upon a severe operation; they may run a low fever, even with sharp rises, or have recurrent or persistent minor upper respiratory infections, etc. While it is clearly folly to operate in the presence of considerable pyrexia or recent increase of illness, we have learnt that it is often better not to wait for seemingly ideal conditions, which may in fact never materialize. It is sometimes best to seize the opportunity offered, for conditions may deteriorate rather than improve.

Moreover, once the anæsthetic or operation has been begun it is desirable, however unfavourable the outlook may appear, to press steadily on until it becomes quite clear that success is impossible. This applies not only to anatomical difficulties but to physiological ones as well. On several occasions the anæsthetist has reported the patient's condition as very grave and the temptation to abandon the operation has been great. In almost all of these cases continuance with the operation after a short wait has been rewarded by final success. The most striking example of the soundness of this policy came in the case of a small child (Case 41) aged 5 years, who was extremely ill; she was deeply cyanosed, with severe recurrent pain in the chest even when resting in bed, and was incapable of any activity at all. During induction of anæsthesia a bronchial spasm developed and she stopped breathing, and it was only with the greatest difficulty that the anæsthetist could inflate her lungs with an intratracheal tube in place; her heart then also stopped. Whether she was then dead is a nice point. Intracardiac injection of adrenalin was given and artificial respiration continued. The heart started again but spontaneous respiration did not begin for another 40 minutes; her condition was, of course, still desperate. At the end of an hour her condition had begun to improve and after much deliberation the operation was started, for it was certain that no second attempt could be made and the outlook was otherwise hopeless. The operation was completed and the child made an excellent recovery; ten days later she was learning to walk about the ward, a thing she had not been able to do before. She has continued to do well.

All our greatest hazards have appeared in the operating theatre; in this series the deaths occurred either in the theatre or within a few hours of return to the ward. If the patient left the theatre in even fair condition, recovery always followed. There was only one instance of bleeding from the anastomosis, that of a man aged 27, very disabled, very blue, who had already had a hemiplegia; he died several hours later from hæmorrhage from the anastomosis which had been quite dry when the chest was closed (Case 48). Bleeding may, of course, occur temporarily when the clamps are

first undone but it either stops spontaneously or has been controlled by insertion of fresh sutures. In one or two of the earlier cases this re-suturing led to narrowing and impairment of the efficiency of the anastomosis and was responsible for some of the poor results.

It has been our experience that patients over 20 years of age carry a far greater operative risk; not only are they commonly severely disabled, but their heart muscle seems to have suffered from the long-continued strain, in contrast to the younger children in whom the myocardium seems surprisingly good. In addition the anatomical hazards may be greater, and the fatal hæmorrhage in this last case was certainly due to the very thin-walled pulmonary artery, aided by a certain degree of extra strain on the anastomosis during systole as the subclavian artery curved down over the prominent aortic arch.

Surgical Problems

Certain general technical features of the operation need discussion in the light of our experiences, for most of these features introduce important practical problems that concern, or should concern, the surgeon undertaking this work.

(*a*) *Blalock's operation or the Potts' modification.* Although the Potts-Smith modification of aortic-pulmonary anastomosis was not used in this series, it has been employed since and has attracted sufficient attention to make it desirable to discuss its advantages and disadvantages. It should be realized that it does not introduce a new principle, but is a technical modification based upon the original principles laid down by Blalock and Taussig. They postulated that certain cases of cyanotic congenital heart disease in whom pulmonary stenosis existed could be improved by anastomosing a systemic artery to a pulmonary artery in order to increase the flow of blood to the lungs, and in their preliminary discussions mentioned the use of the aorta as a possibility in place of one of its branches. It remained for Potts and his colleagues to introduce the ingenious clamp that made this possible.

The advantages of using the aorta would appear to be as follows.

(1) The operation may be easier and quicker.

(2) It is especially useful in small children in whom the subclavian may be too small to furnish an adequate additional blood-flow to the lungs.

(3) It allows the size of the stoma to be varied at will and to be measured exactly.

(4) It may provide a ready solution to the problem of the case with a difficult, deep, short, and narrow subclavian artery.

(5) It avoids the dangers of cerebral damage associated with the use of the carotid or innominate arteries.

(6) It is the simplest, and sometimes the only, way of overcoming the problem of a very high aortic arch with its branches arising at the very root of the neck (Fig. 7).

The disadvantages of using the aorta would seem to be as follows.

(1) It demands a postero-lateral approach (the operation is probably possible, but certainly very awkward through an anterior incision).

(2) It may be very difficult if the aorta is right-sided, as occurs in about one quarter of the cases. This is because the right pulmonary artery is often very short and deeply placed in the mediastinum.

(3) The pulmonary artery may be so small and narrow that it cannot be used, whereas a small thin artery can still sometimes be employed efficiently for end-to-end anastomosis with the subclavian.

(4) In older patients there may be too much disproportion between the size and thickness of the walls of the pulmonary artery and that of the aorta to make a safe junction.

(5) Direct aortic-pulmonary anastomosis may cause a greater strain on the heart and a greater risk of pulmonary œdema.

It is still too early to make a final assessment of the Potts' modification, but the most certain advantages it offers would appear to be a more rapid and less elaborate dissection when the aortic branches are small and deeply placed in the mediastinum, or arise very high in the chest; the possibility of adjusting the size of the stoma to meet the needs of the individual case, especially in very small children; and freedom from the dangers of cerebral damage following use of the carotid or innominate arteries.

In cases where it would be about as easy and satisfactory to use either the subclavian artery or the aorta, it would seem to be surgically sounder and wiser to use the subclavian and to avoid exposing the patient to the greater perils that must attend the deliberate manipulation of a structure of such importance as the aorta. After all, if some mishap befalls the subclavian artery the situation can readily be remedied by ligating it and the aorta could then be used. If some mishap occurs when the aorta is being used it would certainly be more difficult, and perhaps impossible, to retrieve the situation.

(*b*) *Antero-lateral or postero-lateral approach.* Blalock's practice of using the antero-lateral intercostal incision was followed almost exclusively in the first 50 cases, but has often been departed from since then. The antero-lateral thoracotomy incision

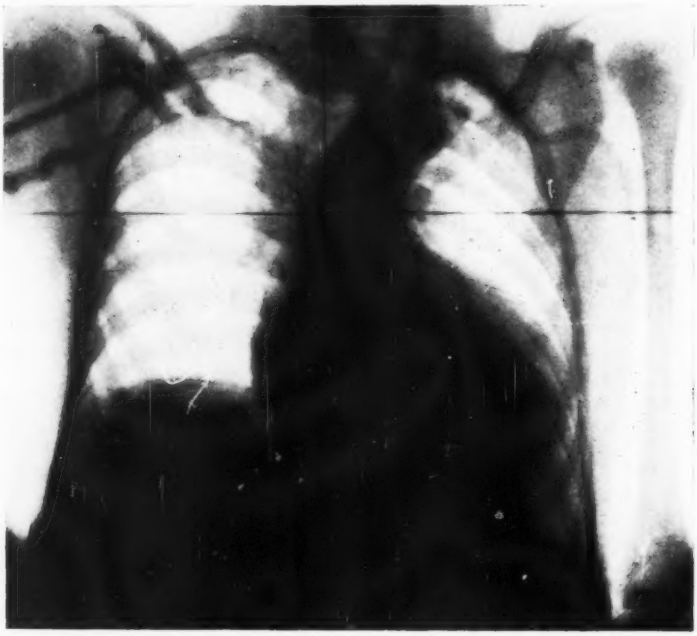


FIG. 7.—Case PO35. Angiocardiogram at 3 seconds. This shows a left-sided aorta which is dilated and very high, with the innominate rising in the ridge of the neck to the right where it had been easily felt in the neck. The illustration also demonstrates the large right to left shunt with pulmonary stenosis.

has always been more popular in American surgery than in that of other countries, as witness the popularity enjoyed by the anterior approach for pneumonectomy and for ligation of the patent ductus arteriosus. In a very ill patient the antero-lateral approach may throw a smaller strain on the lungs and circulation than a postero-lateral one. In general, however, the exposure provided by the antero-lateral intercostal incision may be very cramped, and if the ribs are awkwardly shaped so as to make a high narrow chest, the difficulties of dissection and ligation of the highest branches of the subclavian artery may be extreme. In such cases the faulty exposure commits one to a set of circumstances that really constitute faulty surgery.

One important reason that determined the use of the antero-lateral approach in many cases in this series was our policy of opening the pericardium and examining the heart condition carefully to make as complete a diagnosis as possible and to be prepared to perform a valvulotomy if indicated. The pericardium can, of course, be opened and the heart inspected through a postero-lateral incision, but it would be difficult to utilize the right ventricular approach to the pulmonary valve without considerable and most undesirable dislocation of the heart.

Our present practice is to use the antero-lateral approach only in those cases in which it may be necessary to operate directly upon the right ventricle itself. For all other cases a postero-lateral incision is used with resection of the whole length of the fourth rib from transverse process to costal cartilage. The resection of the rib is much more satisfactory than an intercostal incision which causes more bleeding at the time and may cause dangerous or even fatal oozing afterwards; the risk of bleeding is increased by the use of pericostal sutures, which are also a cause of unnecessarily severe post-operative pain. The postero-lateral thoracotomy with resection of a rib is followed by far less pain than either a postero-lateral or antero-lateral intercostal thoracotomy. The exposure afforded by the long postero-lateral approach is a great advantage and allows a much more easy, rapid, and safer exposure of the vessels and inspires the surgeon with much greater confidence that he has

more complete control of the situation. There is no difficulty in application of the pulmonary artery clamp provided the incision is carried well forwards; the performance of the actual anastomosis is easier than from the front. Moreover, the postero-lateral incision allows use of either the subclavian artery (Blalock's operation) or the aorta (Potts' modification), according to the conditions found.

With but few exceptions, in which the third intercostal space was used, all the operations by the antero-lateral route have been done through the second interspace which is definitely preferable, except in very small children.

(*c*) *Right or left side.* Blalock's earlier recommendation was to use the side opposite to the aortic arch in all but adults and patients in the later teens. His reasons for this were that if the subclavian is used as it arises from the innominate it forms a more satisfactory angle with the parent vessel when it is turned down for the anastomosis; whereas if the subclavian is used as it comes off the aorta it may be sharply kinked at its origin, or flattened as it passes over the prominence of the aortic arch. Also, if the subclavian artery is found to be unsuitable the innominate or the carotid artery can be used instead.

In his latest paper Blalock (1948*b*) states that he uses the right side (when the aorta is left-sided) in all patients between the ages of 2 and 12, but prefers the left approach in children below 2 and in patients over 12 who have attained most of their growth or who are more than 5 feet in height. He mentions that some other surgeons have preferred to use the left-sided approach for all cases.

Blalock's earlier recommendation was followed in most cases in this series, but, recently, departures have been made. In spite of increasing familiarity and experience with the operation the dissection of the systemic arteries on the right side may be extremely difficult, not a little dangerous, and certainly very exacting; especially so when the superior vena cava is large and dilated. It was therefore decided to extend the use of the left-sided approach to children under 12 years as well as to older patients and this was done successfully in a number of cases and with much greater ease; even though the subclavian may appear unduly kinked and flattened at the time the result has been just as good, and it seems probable that the artery elongates and adapts itself. The right-sided approach was then used again on a small child aged 5 years (Case P043) and after a long, tedious, and exacting dissection a very deeply placed and long innominate artery was found which divided high up near the superior thoracic inlet and gave rise to a subclavian artery too short to bring down to meet the right pulmonary artery. This could have been done if the carotid artery had been ligated and divided, but in addition to the carotid another artery almost as large passed into the neck and it seemed that the two vessels must carry a large supply of blood to the head and brain. Alternatively the innominate could have been used, but in addition to the dangers of cerebral ischæmia this artery was so large that there seemed considerable danger of causing heart failure and acute pulmonary œdema if it were used. Accordingly the operation was abandoned with the idea of using a left-sided approach on a later occasion. This experience has finally decided us in favour of using the left-sided approach in all cases unless angiocardiograms suggest the right pulmonary artery is small or absent, indicating it would be dangerous to occlude the left branch while the anastomosis is made. The only other indication for a right-sided approach is the presence of a right aortic arch in a case on which one wishes to do the Potts' operation.

(*d*) *The use of the carotid or innominate arteries.* The danger of cerebral ischæmia is very real if the innominate or carotid arteries are used; in Blalock's series the mortality was 30 per cent. The carotid was divided to allow the innominate to be turned down in two patients in this series (Cases 24 and 35) and in one since (Case P034). The first did extremely well; the second developed an acute pulmonary œdema as soon as clamps were removed and the anastomosis allowed to function, and died after a few hours; the third became comatose and hemiplegic soon after operation and died the next day. Blalock (1948*b*) in his last paper emphasizes that the carotid or innominate should not be used if it can be avoided and states that in many of his earlier cases in which one of them was used, a little longer careful and patient dissection of the subclavian might have spared the carotid or innominate. It would appear to be purely a matter of chance whether interruption of the carotid circulation is followed by paralysis or death or by a good result. It is indeed a gamble, and a poor gamble as well, and therefore is neither surgically nor morally sound. In our opinion, the use of the carotid or innominate arteries is unjustifiable and should be abandoned.

If the surgeon is contemplating using the right-sided approach he should first of all study good angiocardiograms which display the disposition of the aortic arch and its branches. In this way he should be able to assess whether or not it is likely to be possible to use the right subclavian artery. This is a far better way in which to obtain the information than a thoracotomy. An unfavourable

arrangement of the great vessels, such as would make sacrifice of the carotid blood-supply inevitable, is seen in Fig. 7, 8, and 9. In Fig. 7 the aortic arch is unusually high and the innominate artery actually lies above the clavicle and could be seen and felt in the neck; clearly Blalock's operation would be impossible on either side without using a long length of carotid artery from the neck. Such an arrangement seems to demand Potts' operation, which was successfully used in this case. In Fig. 8 and 9 the position of the innominate artery, deep in the mediastinum, is clearly seen and also its very high division with a resultant very short subclavian artery. In these cases the left subclavian artery was successfully used for the anastomosis.

(*e*) *Absent or small pulmonary arteries.* The value of angiocardiography to display the disposition of the aorta and its branches is clearly proven in these cases and it has been of equal value in others. It has been less satisfactory in displaying the main pulmonary artery and its right and left branches, often because of the slow and feeble concentration of the opaque solution in them due to pulmonary stenosis, and especially when a large and rapid shunt has caused the contrast medium to pass rapidly into the systemic circulation. In the normal patient it may be easier to display the chief pulmonary arteries more clearly, especially when oblique or lateral views are used in addition to the postero-anterior ones. One must be prepared to be disappointed with the delineation provided when pulmonary stenosis exists with a large shunt.

The angiocardiograms may, however, indicate that one pulmonary artery is either entirely absent or very small. Fig. 10 shows an example in which the left pulmonary supply seems much smaller than the right. This suggests that it would be unsafe to use the right pulmonary artery as death would probably soon follow its necessary occlusion while the anastomosis is being made. The demonstration or suspicion of a small right pulmonary artery would provide an indication for using a right-sided

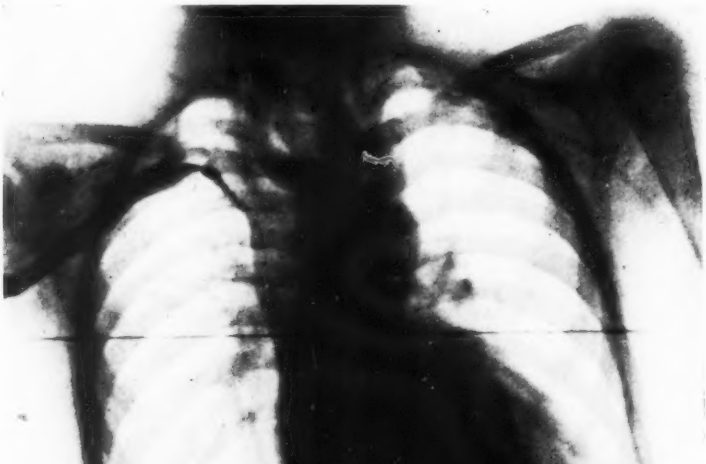


FIG. 8.—Case C017. Angiocardiogram at 3 seconds, showing the origin of the innominate artery fairly deep in the mediastinum with a high division of the subclavian from the innominate and a large gap that would have had to be bridged between this and the pulmonary artery. It also shows an over-riding aorta, a high grade of pulmonary stenosis, and a dilated superior vena cava.

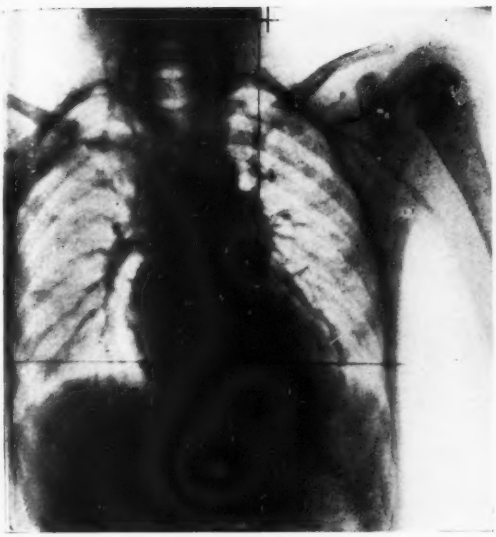


FIG. 9.—Case 0053. Angiocardiogram at 3 seconds. It shows the origin of the innominate artery fairly deep in the mediastinum with a high division and a resultant short subclavian artery that would have been difficult for a pulmonary anastomosis. It also shows an over-riding aorta with moderate pulmonary stenosis.

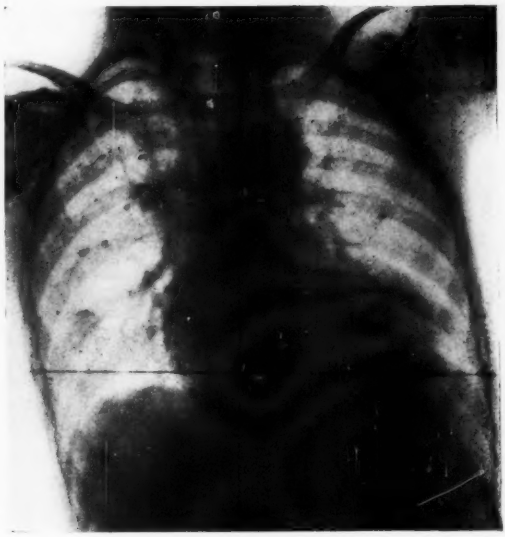


FIG. 10.—Case P048. Angiocardiogram at 4 seconds which suggests that the left pulmonary artery provides a much smaller blood supply than the right and that it would probably be unsafe to occlude the right branch. Operation will, therefore, be performed on the left side.

approach in preference to a left-sided one. Careful screening and plain radiography may also enable one to identify both pulmonary arteries or to suspect that one is small or absent.

In one case, after a long dissection, the right pulmonary artery was found to be not much larger than an intercostal artery and so could not be used; no ill-effects followed the exploration (Case 27). In another (Case 17) the right pulmonary artery was prepared and had been clamped for 12 minutes when the heart stopped; it was assumed that this was due to absence or obliteration of the left pulmonary artery. The heart was started again after massage and injection of adrenalin, and end-to-end anastomosis was performed between the subclavian artery and the first branch to the right upper lobe. Unfortunately the heart stopped several times and finally could not be started again; autopsy confirmed that the left pulmonary artery was completely obliterated. Such cases sometimes are unavoidable and part of the hazards of the procedure, but it may be avoided by more careful radioscopy of the pulmonary arteries.

(*f*) *End-to-end anastomosis.* In order to secure extra length of vessels Blalock not infrequently ligates the pulmonary artery medially, divides it, and performs end-to-end anastomosis to the subclavian. This can be a most valuable step in some of the more difficult cases and may indeed be the only possible way to bridge a gap when the subclavian is short. It is especially useful when, on the left side, the prominence of the aortic arch threatens to kink and flatten the down-turned subclavian artery and to cause tension on the anastomosis. It should certainly be used in preference to end-to-side anastomosis to a narrow pulmonary artery in which most of the width of the pulmonary artery would be encroached upon by the anastomosis. One is naturally reluctant to take the step of ligating and dividing the pulmonary artery but, as Blalock says, a good end-to-end anastomosis is always preferable to an uncertain end-to-side one. The only difficulty may arise from considerable disproportion between the two vessels; Blalock states that he does not mind this provided the pulmonary artery is no more than two to three times the size of the subclavian.

Potts' modification may provide an easy alternative if the subclavian artery is too short to use without end-to-end anastomosis, but if the main

difficulty lies with a small and narrow pulmonary artery, aortic-pulmonary anastomosis may be difficult or impossible and direct end-to-end anastomosis with the subclavian is much the safer and better.

End-to-end anastomosis has been used twice in this series (Cases 8 and 17) and has been used twice since.

Summary of Surgical Procedure

We believe, in the present state of our experience, that the most useful and most satisfactory incision is a left postero-lateral one with resection of the whole length of the fourth rib and incision of the rib bed. This gives a perfect exposure, enabling rapid, comfortable, and much safer dissection of the vessels and also permits use of either the subclavian or the aorta for the anastomosis. We reserve the antero-lateral approach for those cases in which we anticipate that right ventricular cardiotomy may be needed for valvulotomy.

We have abandoned the right-sided approach except for cases in which radioscopy and radiography (including angiocardiography) suggest the right pulmonary artery is unduly small or absent. A right-sided postero-lateral incision is needed if Potts' operation is contemplated in the presence of a right aortic arch.

In general we use Blalock's operation in preference to Potts' modification when it is feasible. The greatest value of Potts' operation is in small children in whom the subclavian artery is too small to furnish an adequate extra flow of blood to the lungs, and in older patients when the subclavian artery is too small or too short and it would otherwise be necessary to use the carotid or innominate arteries. We feel the use of either of these vessels is unjustified, owing to the much higher mortality from cerebral complications or cardiac failure and to the risk of permanent residual paralysis. End-to-end anastomosis of the pulmonary artery and the subclavian artery has a useful place when an end-to-side anastomosis would be difficult, impossible, or under undue tension.

Angiocardiography is an invaluable method to allow pre-operative study and assessment of the pulmonary and systemic arterial pattern.

Much emphasis has been laid on the difficulties and anxieties of these operations, but this gloomy side is relieved by the more satisfactory side of success. In spite of the hazards and long hours of work one derives great satisfaction from contemplation of the successfully completed anastomosis and the rapid, indeed at times dramatic, improvement that follows the operation. As stated elsewhere an excellent result was obtained in 66 per cent; a most gratifying figure when one considers the very poor material with which one is working.

Post-operative Treatment

In our experience the post-operative period is much less stormy and anxious than might be expected. Children particularly tolerate the extensive and prolonged thoracic exploration remarkably well, and the difficulties encountered have been mainly in the older subjects. Penicillin therapy is started 24 hours before operation, and pre-operative instruction in breathing exercises is a routine. Cyanosed and polycythæmic patients should never be left for long periods without fluids and particular care should be given to the fluid intake before operation as a simple precaution against the additional danger of thrombosis at this time.

These patients are well fortified against blood loss during the operation so that transfusion of blood is not needed during or after the operation, but plasma or gum saline are given to combat shock, and the drip is continued on return to the ward. They are encouraged to drink as soon as they recover consciousness. The need for intravenous fluids seldom continues longer than 24 hours, unless the blood pressure fails to rise—it is usually back to the pre-operative level or above in 12 hours—or unless cerebral thrombosis is a complication. An adequate fluid intake of at least 1000 ml. in a small child to 2000 ml. in an adult is needed to prevent thrombosis in the days after operation, and is best given by mouth. With increasing experience we have found that the amount that has had to be given intravenously has decreased; in the first 15 cases it averaged 37 ounces while in subsequent cases it has dropped to an average of 25 ounces. Blood transfusion was needed only in two cases, in one (Case 5) who bled into the pleura and in one (Case 8) who had profound postoperative shock.

The patient is nursed in an oxygen tent on return to the ward and the time that this is needed is judged by tentative periods of removal without development of cyanosis or distress. The time varied from six hours to four days, with an average of 36 hours. Any delay in regaining consciousness or inability to move a limb must be noted as an indication of cerebral thrombosis. Breathing exercises and postural coughing are started as soon as the patient is conscious and it must be emphasized that though this may seem unkind, almost brutal, after a serious operation, the children are not unduly disturbed, and insistence on this early stimulation is amply repaid by the rarity of serious chest complications. A portable radiograph is taken within the first 24 hours and at frequent intervals subsequently, for evidence of collapse and particularly for pleural

effusion. Our experience is that morphine should be used with great care; it is not always needed in the first day, and seldom afterwards. In those with distress from coughing, and this is not common, codeine is useful.

There is an immediate rise of temperature after operation but very seldom above 101°, and excluding four cases with complications where it was prolonged over ten days, the average duration was five days. The pulse rate was the better indication of the general condition: it was frequently at its highest in the second 24 hours, and had usually settled to a steady level of 10 points above the pre-operative level by the end of the first week. The close correlation between temperature and pulse rate, particularly the latter, and the presence of a pleural effusion is mentioned later.

Two indications of the degree of success of the anastomosis in the period immediately after operation are the colour, and the presence of a murmur. There is an immediate improvement in colour at operation as soon as the anastomotic circulation starts, but at this time controlled respiration is occurring. In a successful case this improvement in colour is maintained in the oxygen tent, and after a short period of up to 48 hours will continue outside it. Some cyanosis must be expected from the right to left shunt which remains, and this will be increased if breathing is embarrassed by obstructive secretions in the respiratory tract or by pleural effusions. If these factors are taken into account, a comparison of the colour after operation, particularly of the extremities, with the depth of cyanosis before operation is a reliable guide to the degree of functional improvement that may be expected.

The thrill over the anastomosis, which can be felt by the surgeon, is followed after operation by a murmur heard with the stethoscope; this is usually continuous as heard over a patent ductus arteriosus but may be systolic and diastolic or merely systolic. Though chest complications may make recognition more difficult in the first few days, its presence is an encouraging sign of success and its absence suggests that thrombosis has occurred at the suture line and that the anastomosis is not patent. We have become increasingly impressed with the murmur as a sign that a good result may be expected when the patient is able to start walking, which is usually after the first week.

There are three important post-operative complications: cardiac failure, which is rare; pleural effusion, which is common but generally harmless; and cerebral thrombosis, which is the gravest.

One of the criteria for operation is a heart that can adjust itself to the altered circulation which the anastomosis causes, so that much enlargement of the heart is a contraindication. In this respect the selection of cases would appear to have been satisfactory for there have been few difficulties or anxieties on account of the heart. The blood pressure rises to the pre-operative level, or above, in the first 12 to 24 hours, and any delay in this rise favours the development of thrombosis and calls for the exhibition of a "pressor" drug, such as methedrine, and plasma transfusion. This happened in one of our early cases (Case 8), an adult, where the pressure fell to 50/40 and the systolic was below 100 for the first 36 hours; he developed a cerebellar thrombosis in the first 24 hours. A rise well above the pre-operative level is mentioned by Taussig but we have not experienced it; should it occur, a venesection would be indicated.

It might be expected that the sudden increase in the pulmonary circulation, with the additional work demanded of the left ventricle by the anastomotic shunt induced by the operation, might cause pulmonary œdema in the early post-operative period. We have not experienced this in any case surviving operation though it occurred in one death after operation (Case 35) in one lung on the side where the innominate had been joined to the pulmonary artery. Case 21, a poor result as judged by colour and the absence of a murmur over the site of anastomosis, was slow to recover from this operation, and, despite the absence of a pleural effusion, his temperature failed to settle till the 15th day. On the 27th day he complained of pain over the heart and in the left arm, and was collapsed with ashen cyanosis and dyspnœa; the pulse rose to 140 and the blood pressure dropped to 65 systolic. He recovered with morphia but the attack was repeated next day, and again responded to the same drug. There was no change in the electrocardiograph to suggest a coronary thrombosis. There seems little doubt that these were attacks of left ventricular failure and they were treated as such; he subsequently developed a cerebral abscess which was successfully treated by operation. There were no cardiac incidents in the other patients. Congestive failure was never seen, nor was digitalis used, despite the increased work demanded by the anastomosis, as shown by the increase of heart size after operation in most cases.

The second complication of pleural effusion on the side of the operation is common and occurred in 32 of 43 cases after operation; in 11 it was of moderate size but in 21 it was large enough to demand aspiration, and in 10 of these, more than once. The fluid commonly developed immediately after operation, or in the first few days and naturally it is usually blood-stained. An average example

is shown in Fig. 11. But effusion may suddenly increase, recur, or even first develop later in convalescence and this happened in 5 cases. An early effusion successfully aspirated is on the whole less of a handicap in convalescence than the slowly developing effusion where aspiration is hardly necessary in the first few days and the decision is delayed. It is in these cases that later and repeated tappings are necessary, often difficult, and incomplete in their results. It is these patients particularly whose stay in hospital is prolonged, whose temperature and pulse fail to settle, and 5 such cases were discharged with "pleural thickening" on radiography or showed a very small residual effusion: all cleared up subsequently.

The temperature and pulse chart reflect remarkably clearly the presence of fluid in the chest; an early effusion successfully aspirated is associated with a quick return to normal temperature and pulse rate: a rise on the second and third day, which is not uncommon, almost denotes a slowly developing effusion: a sustained temperature and pulse indicates the persistent effusion, so hampering in convalescence: while with one exception—the case with left ventricular failure—a rise in pulse later in convalescence always pointed to the pleura as the cause. This last point was well shown in Case 39 whose convalescence was extremely satisfactory until the 19th day when he complained of abdominal pain and felt unwell; a rise in pulse rate was the only significant sign but it was sufficient to predict that an effusion was developing on the operative side. It was apparent to clinical signs and radiography the following day and aspiration allowed his convalescence to continue uninterrupted. The late effusion was best illustrated by Case 2 who had a small effusion late in convalescence which resolved, but after returning home a massive effusion developed and necessitated readmission to hospital (Fig. 15, Campbell, 1948). We have regarded these as mechanical setbacks and have not allowed them to retard progress by prolonging bed rest unduly, and this, with the exception of Case 2, has been justified for subsequent examinations have shown a clear and moving diaphragm. There has been no evidence that these effusions are associated with pulmonary emboli and, except once where a very small amount of fluid was noted on the opposite side, the effusions have always been on the side of operation. Persistent hæmorrhagic effusion occurred in Case 5 only, necessitating transfusion; repeated aspiration of 11 pints in all was needed over 4 weeks, the fluid gradually decreasing in colour and gradually in amount until it quite suddenly stopped and did not recur. The correlation between temperature and effusion is roughly shown by an average length of 3 days in those with no fluid, of 5 days in those with moderate effusions, and of 8 days in those where aspiration was needed.

The third and most serious complication of cerebral thrombosis occurred in 3 of the 43 post-operative cases. Case 1, a boy aged 7, with a hæmoglobin of 126 per cent, had a history of a brain abscess on the left side when 2 years old. A right hemiplegia of moderate severity which was noted immediately after operation began to improve on the seventh day and when he was discharged on the sixteenth day there was only a slight limp. He has made a complete recovery. Case 8, an adult, was a severe case with hæmoglobin of 141 per cent and had the severe drop in blood pressure in the first 36 hours already noted. The two factors of marked polycythæmia and shock were therefore present to encourage thrombosis, and this was noticed in the first 24 hours, the left anterior cerebellar artery being involved. Recovery from this was complete but on the eighth and twenth-fourth days he had thrombosis of systemic arteries and on the twentieth day a thrombosis of the anterior cerebral artery. From this severe complication he has made a gradual but not a complete recovery, a disappointment in view of the excellent physical result. The third, Case 29, was a boy of ten, with a hæmoglobin of 158 per cent; he was drowsy after operation and was found to have a thrombosis of the cortical ascending parietal branch of the left middle cerebral affecting the arm area. The blood pressure was not unduly low after operation but during the operation there was marked collapse when the right pulmonary artery was occluded, and it was probably then that the thrombosis occurred. He is left with impairment of movements of the right arm and hand, a considerable disadvantage though compensated by an otherwise excellent result.

A further severe complication which we regard as due to the operation was in Case 21 who was admitted to Oldchurch County Hospital one month after discharge from Guy's after a prolonged post-operative stay of 56 days due to persistent temperature and left ventricular failure as already mentioned. The cause of this was a cerebral abscess which was successfully dealt with by aspiration one month later. It is not impossible that a post-operative thrombosis was the basis of this complication, though it was some time after.

It will be seen that cerebral thrombosis is the most important complication and one which to a lesser or greater degree nullifies the physical benefits that an otherwise successful operation gives. It should be most feared in those with gross polycythæmia.

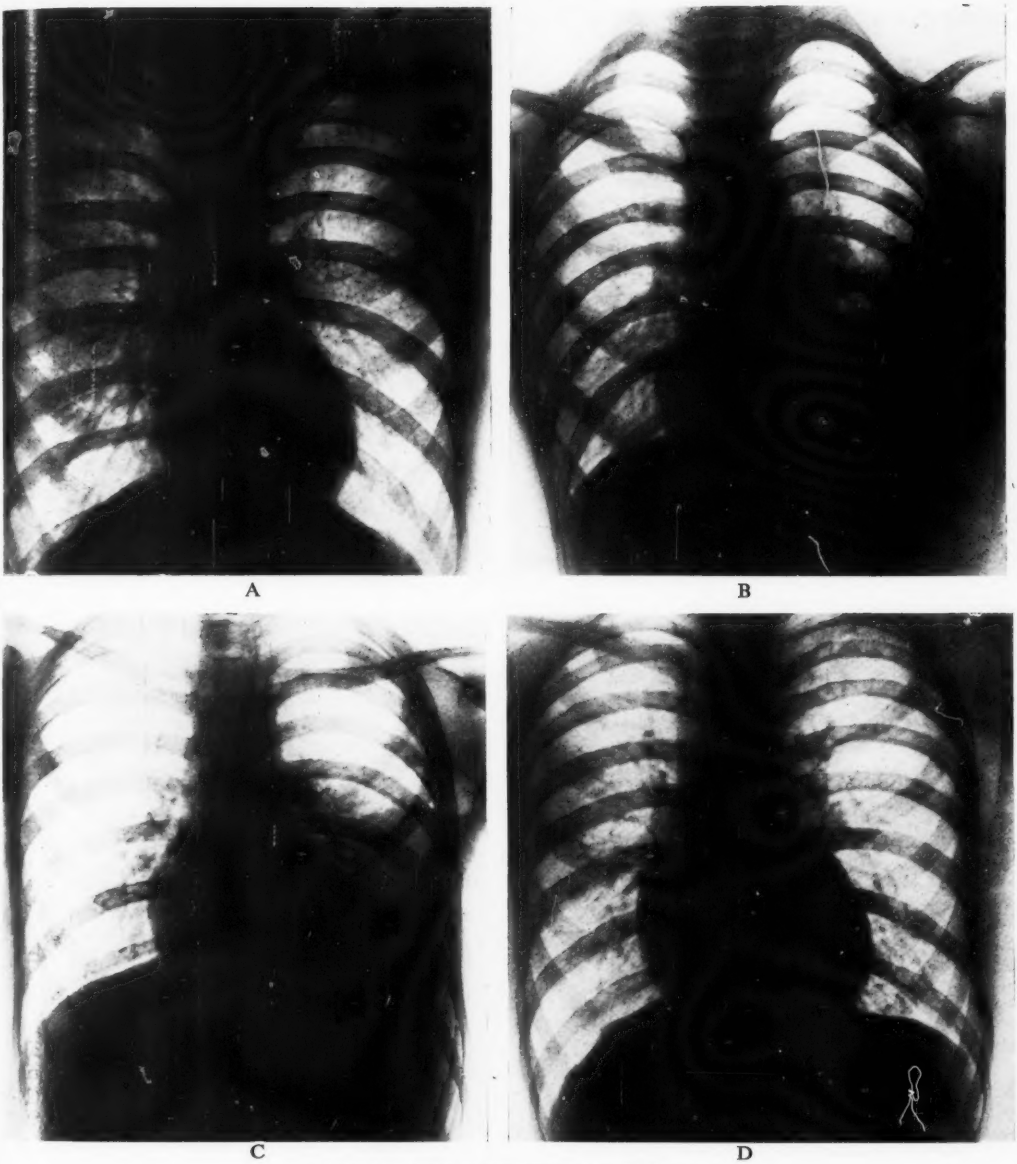


FIG. 11.—Case 26 (Reference No. P069).

(A) Teleradiogram in P.A. position before operation showing the clear lungs and some pulmonary bay with a raised apex. Both pulmonary branches can be seen (9/1/48).

(B) Portable radiogram, one day after operation, showing a moderate sized effusion at the left base with some collapse (11/2/48).

(C) Portable radiogram, showing gradual clearing of the effusion, the heart size becoming more visible but not yet easy to measure accurately.

(D) Teleradiogram, 7 months after operation, showing some increase in heart size from before operation, c.t.r. 54 instead of 46, with less apparent change in the density of the lung than in many cases, although the result was most successful.

with severe operative and post-operative shock, and in adults more than children. Prophylaxis with anti-coagulants introduces the danger of hæmorrhage and for this reason we do not use it. Nor have we felt sufficiently confident to start treatment by heparin within three days of operation, though there is a strong case for giving it promptly if thrombosis is diagnosed. If blood loss at operation has not been great, and polycythæmia is still present, blood letting is indicated and also intravenous fluids, by which method the heparin can be given.

In comparison with these three, other post-operative complications are slight. Some collapse of the lung on the side of operation, independent of effusion, occurred and required no treatment except continuation and insistence on breathing exercises. A small pneumothorax was present in one case and required no treatment and in only one was surgical emphysema of any degree present. Obstruction to breathing by secretions in the upper respiratory tract is largely obviated by insistence on breathing exercises and postural coughing, but was present in 17, though in only 4 did it cause any anxiety. Case 3 responded to the old fashioned steam tent, and Case 49, after 36 hours of obstructed breathing, to the more old fashioned method of turning him upside down, both exhibited by a watchful and wise ward-sister. Cases 40 and 41 both had severe laryngitis, and in the first, where a heavy growth of yeasts was cultivated from a throat swab, tracheotomy was necessary but led to little delay in his complete recovery.

The disability in the arm on the operated side is remarkably slight, and though the vascular change is apparent in signs, symptoms are slight and the disinclination to move it is soon overcome. A diminution in the size of the pupil on the side of operation, occasionally with ptosis, is almost a constant finding but had generally disappeared by the time the patient was discharged. No disturbance in kidney function was noted.

On the whole the postoperative period is surprisingly tranquil and uneventful for such an extensive and eventful operation. Children stand it particularly well and it is the adolescent or adult who is more likely to present difficulty. No better example of this can be shown than Case 41, a gravely ill girl of 5, whose heart, as well as breathing, stopped during induction of anæsthesia. Despite this and an operation lasting over three hours, and later a severe tracheitis, she was out of bed on the fourth day. Excluding 12 cases where the stay was prolonged on account of cerebral thrombosis, wound sepsis, or pleural effusion, the average stay in hospital was 23 days, and the average time in bed was only 7 days.

Results of Operation

In these first 50 patients there have been 7 deaths, a mortality of 14 per cent. Two of these deaths were among the relatively small number of patients of 19 and over (Cases 1A and 48). One had a single pulmonary artery and died when it was clamped (Case 17). In the others, there seemed no special reason except their serious condition and the severity of the operation (Cases 12, 32, 35, and 38).

Fallot's tetralogy was present in five of the seven. The remaining two had somewhat more complicated forms of morbus cœruleus, one having a single auricle in addition (Case 35) and the other having an infundibular stenosis, an aorta arising from the right ventricle without transposition of the pulmonary artery, and a small left ventricle with its only exit a ventricular septal defect (Case 32).

There were three patients where no anastomosis could be performed. One, an error in diagnosis already described, because the pressure was high in the pulmonary artery (Case 28), one because the right pulmonary artery was too small (Case 27), and one because of the technical difficulties introduced by the enormous dilatation of the collateral circulation (Case 14). In no case was it impossible to find a suitable systemic artery, but often the shortness of the subclavian led to practical difficulties and was, we think, sometimes the reason for the lack of success. This leaves 40 cases to be considered.

There were three where, in our opinion, there was no improvement (Cases 11, 19, and 22), though even then the parents of two thought there had been some. In one, the anastomosis was thought to be too small at the time (Case 11) and in a second there was bleeding from the anastomosis when it was nearly completed, and to save the patient's life many stitches had to be put in that probably led to some intra-arterial thrombosis and occlusion of the anastomosis (Case 22). It is possible that these three were also errors in diagnosis and not technical failures of the operation, but we do not think so.

In four the result has only been included as "fair." In one of these a similar difficulty with bleeding from the anastomosis occurred (Case 43); after operation he was able to walk upstairs which he had never done before but was not greatly improved. In the second (Case 21) the result was at first classed as a failure and after a long convalescence a cerebral abscess followed: when seen later after this had been cured, he was certainly improved but not as much as most of the others. In the other two the result was as good as possible as far as the heart was concerned, but the patients were handicapped by thrombosis resulting in partial hemiplegia; in one of these severely (Case 8), and

in the other, causing much limitation of his arm and hand movements but not preventing a great increase in his physical activity, so that he could walk about the greater part of the day (Case 29).

This leaves 33 of the 50 cases (66 per cent) where the result was almost perfect. The patients' capacity was enormously increased up to walking several miles instead of a few hundred yards, or getting about all day instead of being an invalid mainly in the house. To the parents the improvement seemed almost miraculous. One boy (Case 1) who had been tied to his mother's apron strings was at school within two months and after a year was running about all day, playing cricket, and could easily walk 4 miles. After running fast for 25 yards there was slight cyanosis in his lips and nails and some dyspnœa, but this cleared up very rapidly.

Another (Case 2) who had rarely been out of his parents' sight had within a year walked 6 miles and climbed hills in Switzerland as easily as his parents: he was anxious to become a medical student and was doing well at school.

Another (Case 4), who had been carried into hospital by his father, was leading a normal life at school and was able to go roller skating within a few months. Case 23, aged 19, who had been a complete invalid, had been on a camping holiday and had walked 6 miles. Such accounts could be repeated for nearly all the patients and all who have done less well have been mentioned individually.

The cyanosis was also much improved. In most of them it was absent on casual inspection though it could generally be seen in the nails at rest (but often not in the lips) and tended to be noticeable only on a cold day or after vigorous exertion. In one of the severe cases (Case 6) the cyanosis was still moderate, but it had been extreme and the child was able to do so much more that we feel it right to include her in this group. Case 49, with tricuspid atresia, who was very severely limited has not yet made as much progress as the others, but after leaving hospital fit and apyrexial, he was admitted elsewhere a month later with suspected bacterial endocarditis. He made a good recovery and can get about all day and his limitations seem more his unusually poor muscles than his heart. His colour has improved, but cyanosis can still be seen in his lips. We hope on the analogy of other cases that his improvement will go still further as his muscles improve.

The clubbing of the fingers also disappeared in some patients in the course of four or five months; some improvement was generally noticed very early. Where the clubbing was gross it seems more doubtful if it will disappear entirely, though it has changed greatly and become much less noticeable in the course of six months. No trouble has been experienced from the arm where the subclavian was divided, though the brachial pulse has not returned with sufficient strength to be able to measure the diastolic blood pressure in this arm.

In nearly all the successful cases a continuous murmur, such as is heard with a patent ductus, has persisted though often the thrill seems less than might be expected with the murmur. This is a good guide to the success of the operation and unless such a murmur can be heard early and easily, it is unlikely that the patient is going to be one of the most successful results. Occasionally, when there was less improvement than usual, there was a systolic murmur only.

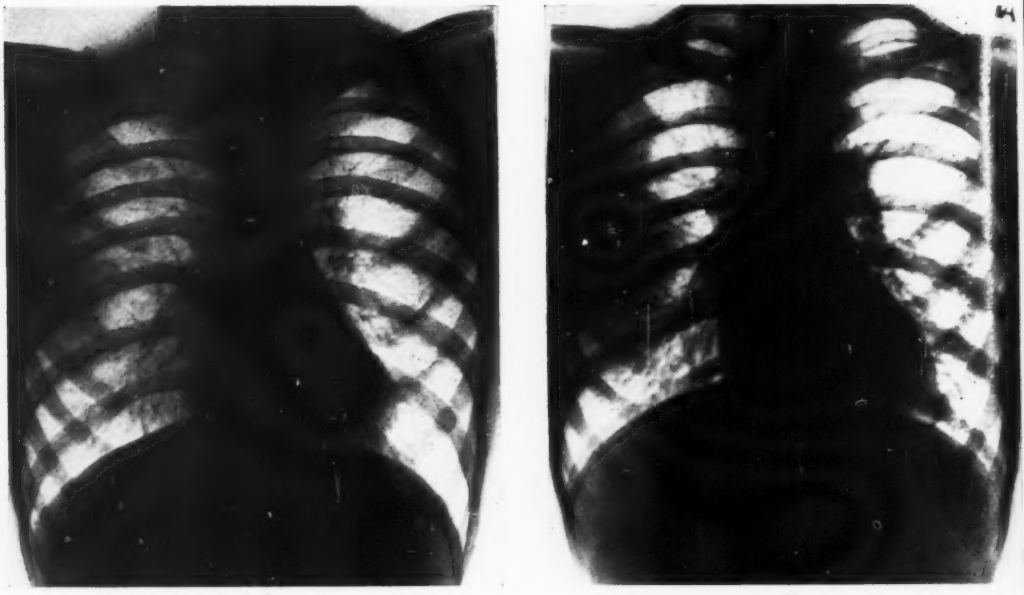
The enormously increased ability of these patients to get about often reveals orthopædic disabilities that have not mattered previously. Owing to the poor development of the muscles through lack of use and the small blood supply, and sometimes to an added deformity of the limbs from their continuous squatting, their new activity reveals many postural defects. We have made it almost a routine for the patient to have exercises and to be trained in walking correctly, and with simple supervision on these lines and occasionally with wedging of the shoes they have made rapid progress and had no serious difficulties.

We were at first a little surprised that there was not a quicker increase in the weight. As already stated, parents were nearly always anxious about the difficulty of making their children eat, and generally the appetite improves at once and they eat well. Probably the greatly increased activity prevents them having anything to spare for putting on weight for a time, but they can be expected to gain after a few months rather than a few weeks, and most of them have increased and gone some way to catch up with the weight that is normal for their age.

Increase in Size of the Heart

One of the points that has been emphasized as a drawback to the Blalock-Taussig operation is the increase in the size of the heart that may be expected from the work added by the left to right shunt. We have, therefore, paid special attention to the size of the heart before and after operation.

Of the 50 cases, 7 died, 3 had no anastomosis, and in 3 the operation was thought to have been unsuccessful. This leaves 37 where the heart size can be compared before and after a successful anastomosis. Owing to many of these patients living so far from London it has not been easy to get a regular follow up. We have, however, seen and obtained reasonably comparable radiograms in all



A B

FIG. 12.—Case 23, a boy aged 19, with Fallot's tetralogy.

(A) Before operation, showing a slightly raised apex and a straight left border.

(B) Eight months after operation, showing some increase in the size of the heart, the c.t.r. being 48 instead of 42, but still within normal limits. There is also some increase in the shadow of the pulmonary artery, especially on the left, the side of the operation as the aortic arch was right-sided.

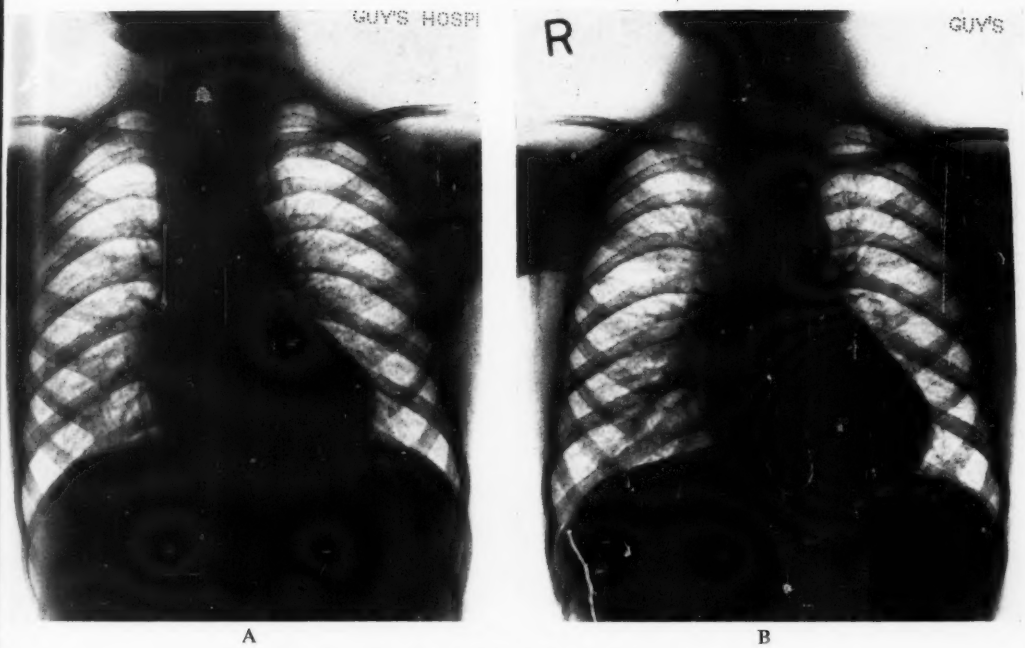
but two where we are relying on reports from their doctors: in neither of these had the heart increased in size a few weeks after operation. In one patient who lived in Cyprus we were fortunate in getting a report and teleradiogram from Dr. Hills who had taken the films at Guy's Hospital before operation. Examples of an increase of average amount, of very little change, and of the greatest increase we have seen are given in Fig. 12, 13, and 14 respectively.

This leaves 35 cases to be considered, and we have taken separately those where the follow up was more than a year after the operation.

Of the cases operated on more than a year ago all 13 have been followed up and teleradiograms have been obtained. Two of these showed no significant change in the size of the heart, but there was some increase in the other 11. In 6 cases there was no further increase after the first month and in another 3 there was little or no increase after about four months, but 2 who had not shown much increase in the first few months showed some increase between four months and a year. The average increases, shown in Table III might look like a slow but progressive rise; however, consideration of the individual cases shows only 3 out of 13 with any increase after four months (from 51 to 55). Naturally these three will be followed up with special interest to see if they are in fact exceptions.

Of the remaining 22 cases, some had been followed up for 11 months and all for more than 7 months, except three so far only followed for 5 months. Three of these showed no significant change in the size of the heart, but there was some increase in the other 19. In 10 cases there was no further increase after the first 4 to 6 weeks; in 9 there was some increase after this, but in 4 of the 9 it was only by 2 or 3 points per cent and in another 3 by only 4 points per cent. We had not, as a rule, intermediate records at about 4 months in these patients, but in the three where we had, the increase had all taken place by this time. The average increases are shown in Table III.

Taking the two groups of cases together the average increase in cardio-thoracic ratio was from 48·0 to 52·7 per cent. It had not increased in 5 of



A B

FIG. 13.—Case 29, a boy aged 10, with Fallot's tetralogy.

(A) Before operation showing a blunt apex and a fairly large pulmonary bay.

(B) Four months after operation showing hardly any increase in the size of the heart (c.t.r. inchanged at 52) although the anastomosis was most successful, as shown by his improved ability to get about and the improved colour. The pulmonary artery is more prominent on the right which was the side of the anastomosis as the aortic arch was left-sided.

TABLE III

CARDIO-THORACIC RATIO BEFORE AND AFTER BLALOCK-TAUSSIG OPERATION

Number of cases	Before operation	After operation			Time after operation
		About one month	About four months	At last visit	
13	47·2	51·0	51·8	52·5	12 mo.
22	48·6	50·9	—	52·8	6–11 mo.

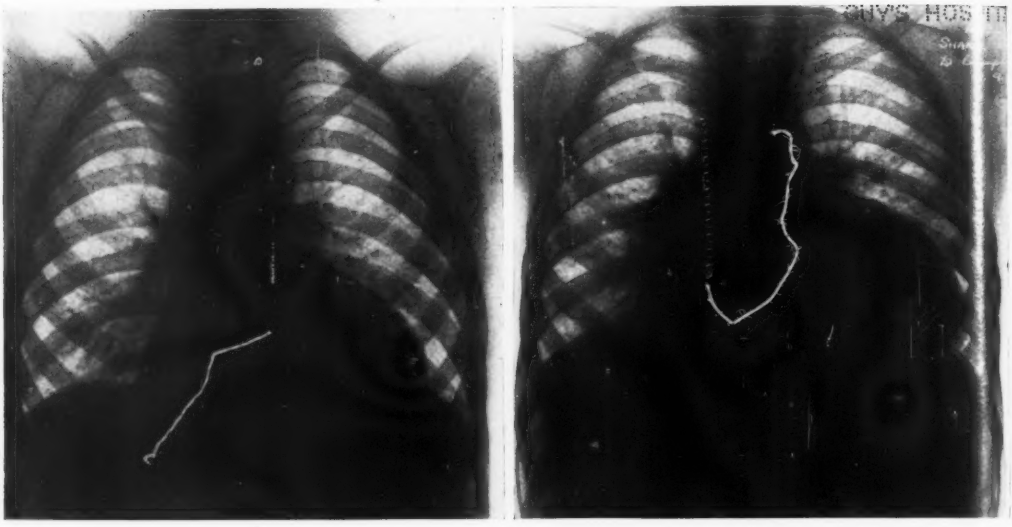
t e 35 cases. It had increased by 2 to 3 points per c nt (from 48 to 50 or 51) in 8, by 4 or 5 points per c nt in 10, by 6 to 8 points per cent in 10, by 10 points per cent in 1, and by 12 points per cent in 1 case. In the last two the hearts had been small before operation (c.t.r. 41 and 44).

Only one patient (Case 33, Fig. 14) has made us anxious about his future by the degree of increase in the size of his heart. His clinical improvement was as much as in the others and there was difficulty in restraining him from doing everything. He was treated as a normal child at school except for games and had nothing except occasional attacks of tachycardia.*

PULMONARY VALVULOTOMY

During the same period 6 patients have been submitted to the operation of pulmonary valvulotomy. Four of these were thought to have Fallot's tetralogy with pulmonary stenosis as an important feature and the other two were thought to have pure pulmonary stenosis with some degree of patent

* We have since heard that eight months after operation he developed bronchitis with a temperature of 103 degrees; œdema of the legs soon followed, the temperature persisted, and he died in a few days.



A B

FIG. 14.—Case 33. From a boy aged 4, with tricuspid atresia and non-functioning right ventricle.

(A) Before operation showing an enlarged and rather horizontal heart.

(B) Five months after operation illustrating the greatest increase of heart size seen so far in any patient after operation, the c.t.r. having increased from 61 (11·0/18) to 69 (13·0/18·75). The pulmonary artery had become more prominent on the right, the side of the anastomosis, as the aortic arch was left-sided.

foramen ovale or auricular septal defect. The operation appears to be of much greater danger and three (Cases P031, H117, and H121) of these six have died, but they were all older patients, 22, 24, and 29, and this and the relatively small numbers make any exact comparison impossible.

In the remaining three, the operation was successful and the patients were considerably improved in colour and in their ability to move about without acute dyspnœa. Unfortunately, two of these were handicapped, one by hemiplegia (Case H109) and one by some disability of the leg from arterial embolism (Case P079). Whether this is a greater risk with valvulotomy remains to be seen because here again, they were older cases, aged 18 and 23, and it appears that age (though it may be the associated degree of polycythæmia) makes thrombosis and embolism a greater risk.

The remaining patient (Case H107, Case 2, Brock, 1948), a girl aged 11, was as brilliantly successful as any with an anastomosis. She was active all day and was able to walk several miles and looked a normal colour with only trivial or even doubtful cyanosis on careful examination. Her arterial O_2 saturation had increased from 81 to 91 per cent. Twelve months after operation, she has developed no signs suggestive of pulmonary regurgitation.

SUMMARY AND CONCLUSIONS

The method of chosing the first 50 patients for the Blalock-Taussig operation at Guy's Hospital and the results obtained have been described. Three cases were thought to have tricuspid atresia with a non-functioning right ventricle, and all the others Fallot's tetralogy, though sometimes with a known or suspected complication. All had great disability and severe or moderate cyanosis dating from birth or early infancy, with polycythæmia and clubbing of the fingers.

Most of these patients had a systolic murmur—often not very loud—in the pulmonary area, and in about half of them a thrill could be felt at this site. None had a diastolic murmur or a greatly accentuated second sound. Four-fifths of the patients squatted habitually and panting on exertion was nearly as characteristic.

The heart was generally within normal limits though some right ventricular hypertrophy could be seen on cardioscopy and shown electrocardiographically; sometimes the heart was small, and

occasionally a little, but never much, enlarged. If the cardio-thoracic ratio is under 45, the heart will probably stand a fairly large anastomosis with still greater improvement for the patient. More experience will be needed to decide how much enlargement of the heart may be allowed: slight enlargement with the c.t.r. 52 to 54 should certainly not contra-indicate operation but larger hearts with the c.t.r. 55 and above require special consideration and the improvement may not be so lasting.

The heart was sometimes sabot-shaped and sometimes of more normal shape with a straighter left border or even with a slight prominence of the pulmonary region. The pulmonary vascular shadows were generally much diminished though sometimes more mottled shadows, probably produced by the collateral circulation, made this decision difficult.

The pre-requisite of a successful operation is that the disability and cyanosis should be mainly due to an inadequate blood flow to the lungs and, in general, this is indicated by the clinical and radiological findings that have been given.

The other prerequisites are that there should be a suitable systemic artery and a pulmonary artery large enough for an adequate anastomosis. Angiocardiography helps with both these points by showing the anatomy of the aortic branches and of the pulmonary artery and so in helping one to plan the details of the operation that is most likely to be feasible and successful. Careful radioscopy should generally be able to decide about a suitable pulmonary artery.

The operation was usually an end-to-side subclavian-pulmonary anastomosis on the side opposite to the aortic arch. The aortic arch was right-sided in a quarter. Reasons have been given for thinking it is better to operate on the left side regardless of the side of the aortic arch and this has become the usual routine recently. Our conclusions about the surgical procedure that should be adopted have been summarized (page 188).

The immediate upset caused by the operation is less than might be expected and children stand it well and recover quickly. Morphia should be used in small quantities only, and intravenous fluids are not needed in large amounts, though fluids should be taken freely by mouth before and after operation. Breathing exercises should be started at once and carried out vigorously. The only complications that are at all common are pulmonary—some collapse of the lung and pleural effusion. These generally clear up quickly though they often need aspiration once and may sometimes be slower and cause trouble. Arterial thrombosis at the time of operation or soon after has been the second main complication and as already stated two of these patients have been left with some residual disability. This risk seems greater when the polycythæmia is severe.

On the average the temperature had settled to normal by the third day, and excluding 12 cases where there were complications from cerebral thrombosis, wound sepsis, or pleural effusion, the patient was up and getting about the ward on the eighth day and was able to leave hospital after 23 days.

There were 7 deaths in these 50 cases—a mortality of 14 per cent. There were 3 others where no anastomosis could be performed.

Of the remaining 40, there were 3 where we thought the improvement of little or no significance, and 4 where we have only classed it as fair, though in 2 of these the result was excellent as far as the heart was concerned but was marred by some residual disability from thrombotic hemiplegia.

In 33 of the 50 cases (66 per cent) the results were almost perfect and the patient was able to get about all day and walk up to 5 or 6 miles instead of a few hundred yards or less. The cyanosis almost disappeared except slightly in the nails and on a cold day. Many children quickly started at ordinary schools, and cricket, camping, and roller skating became the occupations of some who had before been invalids doing hardly anything.

A murmur similar to that of a patent ductus arteriosus was heard on the side of the operation in the successful cases. After operation with the added work of the heart from the new left to right shunt the heart generally increased in size a little but not greatly. In 5 of 35 cases there was no significant increase; in the remainder the cardio-thoracic ratio rose from 48·0 per cent to 50·9 when the patient left hospital and to 52·7 per cent when they were last seen (generally after 7 to 14 months). Although these average figures might look like a progressive, if slow, increase, individual cases show that the greater part of the increase had generally taken place in the first 4 to 6 weeks. Only one patient so far—and he had tricuspid atresia—has caused us any anxiety for the future by the size of the increase in his heart, and symptomatically he has improved as much as the others.

It was particularly encouraging that none of the patients seen a year after operation had failed to maintain or increase all the improvement they had made at the earlier follow-up after a few months.

We would like to thank Dr. T. H. Hills for the angiocardiograms and the Photographic Dept., Guy's Hospital for help with the illustrations.

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Appendix on Cardiac Catheterization

By G. A. Zak

Nine of these cases underwent cardiac catheterization. Most were investigated when the procedure was still at an early stage of development at this hospital; with increasing skill and improved technique the results in later cases have been more complete.

In view of the envisaged operation the main object of venous catheterization was to get information regarding the pressure in the right ventricle and pulmonary artery, the presence and localization of shunts, and the volume of the pulmonary blood flow in relation to the height and weight of the patient.

It is not intended to discuss the problems and findings of cardiac catheterization in detail. From the rapidly growing data on this subject several widely accepted facts have emerged. For example the presence of blood of significantly higher oxygen saturation in the right auricle than in the venæ cavæ indicates arterial blood passing from the left auricle through an auricular septal defect; and the sudden fall in pressure from a high to a low level on passing from the right ventricle to the pulmonary artery is the best evidence of pulmonary stenosis. To calculate the pulmonary arterial flow with the aid of the Fick principle mixed venous blood must be obtained, and the mixing of the venous blood from the various sources may be incomplete in the right auricle. If the catheter can not be got into the pulmonary artery a blood sample obtained from the outflow tract of the right ventricle can be substituted. The pulmonary arterial flow is not necessarily equal to the total pulmonary flow (pulmonary capillary flow) as an appreciable amount of blood may reach the alveolar capillaries by way of a collateral circulation. No attempt has been made to assess this.

The sudden fall from high to low pressure on passing from the right ventricle to the pulmonary artery and sometimes the feel of release from a grip on the catheter is the best evidence of pulmonary stenosis, and a decreased pulmonary arterial flow is confirmatory evidence. If it is not possible to reach the pulmonary artery with the catheter, a high pressure in the right ventricle and a decreased pulmonary flow may be taken to lend good support to an assumption of pulmonary stenosis in the absence of clinical signs of mitral stenosis or heart failure.

The magnitude and direction of an overall intracardiac shunt is calculated by deducting the pulmonary arterial from the systemic blood flow or vice versa. The adjective "overall" is used to denote that in many instances there is actually a to-and-fro movement of blood from the two sides across a single septal defect but as a rule one component is larger to such an extent as to dominate the clinical picture.

An evaluation of the output of each of the two ventricles of the heart, and the consequent calculation of a possible shunt is an approximation only. The calculation of the pulmonary arterial flow is often the more reliable. Of the three components necessary for its computation, the O_2 uptake through the lungs and the O_2 content of the mixed venous blood are known. If the degree of the arterial oxygen saturation is normal the pulmonary venous blood is taken to be of equal saturation. Even if it is lowered one is justified in assuming the pulmonary venous blood as still being 95 to 98 per cent saturated with O_2. This was shown to be true by other investigators in the absence of pulmonary conditions likely to interfere with the gaseous exchange in the alveoli.

Case No., Age, and Date of catheterization	Pressure (mean) in mm. Hg. from the skin of the back			Systolic B.P. (clinically)	Degree of O_2 saturation percentages				Output in % of the average output of 3·0 l. per min. per sq. m. of body surface		Hb. % 100% = 15·6 g. Hb.	Surface area in sq. m.	O_2 consumption in ccs. per min. at S.T.P. (B.M.R.)
	R.A.	R.V.	P.A.		R.A.	R.V.	P.A.	Syst. artery	System	Pulm.			
Case 23, 19 yr., 13/1/48.	15	43	—	115	29·0	33·1	—	46·5	100	30	120	1·24	170 (−5·0)
Case 49*, 8 yr., 6/2/48.	14	(27)	—	100	SVC 46·8	23·2	—	58·8	180	—	125	0·83	140 (−3·0)
Case 35, 7 yr., 23/4/48.	7	45	15	110	—	—	—	—	—	—	—	—	—
Case 36, 17 yr., 12/5/48.	12	—	—	122	60·0	—	—	83·0	85	—	118	1·42	200 (0·0)
Case 42*, 19 yr., 20/5/48.	11	—	—	95	55·7	—	—	73·0	110	—	140	1·32	220 (+19·0)
Case 39, 11 yr., 25/5/48.	16	44	—	100	47·7	—	—	55·6	250	—	143	1·00	185 (+4·5)
Case 48, 27 yr., 4/6/48.	15	50	—	115	38·2	46·6	—	62·7	90	35	153	1·77	250 (+4·0)
Case 44, 11 yr., 15/6/48.	10	40	10	110	67·6	60·7	60·1	71·8	110	45	160	1·01	165 (−5·5)
Case 46, 15 yr., 18/6/48.	10	62	10	125	53·3	55·4	46·6	61·8	100	50	142	1·59	210 (−5·5)

* In these two cases the diagnosis was tricuspid atresia; in all the others it was Fallot's tetralogy.

The systemic blood flow in cases of septal defects cannot, however, always be calculated with the same degree of reliability as the right ventricular output. Here, the mixed venous blood component is the weak link in the chain. Often there is evidence of imperfect mixing in the right auricle, the coronary vein as a rule adding blood very much lower in oxygen content. The inferior vena cava in turn produces, generally speaking, more saturated blood than the superior vena cava. A blood specimen obtained from the inferior vena cava, however, may not be of uniform composition on account of the nearness of the openings of the hepatic veins. This fact makes the recognition of small atrial septal defects producing admixture of arterialized blood uncertain if one is to rely on the taking of single blood samples only. Because of the errors involved in calculating the pulmonary and the systemic flow one must hesitate to diagnose a small overall right to left shunt on small differences; larger differences can however be taken as reliable.

In these nine cases the clinical diagnosis was supported. Such support was felt to be forthcoming in cases of Fallot's tetralogy if there was evidence of pulmonary stenosis and an overall right to left intracardiac shunt.

Of the two cases diagnosed on clinical grounds as having tricuspid atresia only a single right chamber with normal right auricular pressure could be demonstrated and no evidence of an atrial left to right shunt were found. In neither of these cases could the catheter be made to cross over to the left auricle.

There was evidence of an overall right to left shunt in every case.

The A–P diameters of the chest ranged from 18 to 24 cm. with an average of 19 cm. To obtain the pressure as measured from the sternal level 15 mm. of Hg. has to be deducted from the values given in this table.

The catheterizations were carried out by Dr. G. A. Zak and Dr. H. E. Holling.

Case 23. Pulmonary artery not entered as tip of catheter got caught in papillary muscles towards the base of the heart, without getting near the pulmonary valves.

Case 49. Results vitiated by crying. The superior vena cava value is substituted for the right auricular value, as the former followed a period of calmness. It was thought at first that the right ventricle had been entered, but probably the catheter was in the coronary sinus, with the pressure high from partial occlusion of the lumen of the catheter.

Case 35. The use of an inhalation anæsthetic made gas analyses of the blood specimens impossible. The raised right ventricular pressure in conjunction with the lowered pulmonary arterial pressure indicates stenosis of the pulmonary ostium.

Case 36. Venospasm led to abandonment of procedure.

Case 42. Only the right auricle could be entered. The tip did not pass into the left auricle through a possible atrial septal defect. If such a defect was present, a left to right shunt through it would appear unlikely on account of the S.V.C. and I.V.C. saturations, which agreed with the value found in the R.A.

Case 39. The right ventricle was only entered with difficulty and attempts at entering the pulmonary artery failed. Tip was at the base of the heart. The pressure in the right ventricle was recorded but no blood specimen was obtained, and when on withdrawing the catheter too far this chamber had been left the permissible screening time did not allow further search for the right ventricle.

Case 48. The pulmonary artery could not be entered though tip of catheter was brought to base of heart. Good agreement between S.V.C. and right auricular saturation. The increased right ventricular saturation over the right auricular sample favours presence of a ventricular septal defect, giving rise to shunting of the blood in both directions though differing in quantity. The smaller opposing shunt can be detected if the tip of the catheter happens to be near such a defect, which seems to have been the case here.

Case 44. Superior vena cava, 54 per cent, and inferior vena cava, 60 per cent saturated, favour a small left to right shunt through an atrial septal defect, which is, however, far outweighed by the large right to left shunt through a ventricular septal defect or overriding of the aorta.

Case 46. Good agreement between the saturation values in both venæ cavæ, right auricle, and right ventricle. The low pulmonary artery saturation is thought to be due to obstruction by the catheter of an already narrowed ostium for about two minutes, prior to withdrawing the blood sample. The pulmonary flow has been calculated from the right ventricular saturation.

ABSTRACTS OF CARDIOLOGY

The Electrocardiogram in Biliary Tract Disease and During Experimental Biliary Distension. Clinical Observations on 26 Patients. G. B. Hodge and A. L. Messer. *Surg. Gynec. Obstet.*, **86**, 617–626, May, 1948.

The effect on the electrocardiogram of experimental distension of the biliary tract was investigated in 26 patients undergoing surgery of the biliary tract. Sterile normal saline solution was introduced under pressure through a cannula into the gall-bladder or through a rubber T-tube into the common bile duct; the maximum pressure used was 100 cm. of water. Twenty-two of the patients had chronic cholecystitis, 3 had previously had cholecystectomy, and 1 had a carcinoma of the head of the pancreas with chronic cholecystitis and cholelithiasis. In 14 patients gall-bladder distension and electrocardiographic studies were carried out simultaneously during operation. Of a second group of 13 patients the common bile duct was distended in 12 and the gall-bladder in 1 patient, without medication or anæsthesia, 10 or more days after operation. No patient had angina or myocardial infarction and in none did distension of the common duct or gall-bladder cause anginal pain. All patients who experienced pain during distension of the common duct or gall-bladder complained of respiratory distress during distension and in the majority the blood pressure rose. No constant cardiographic changes were found as a result of the distension; control records obtained before operation included abnormal as well as normal tracings. It is concluded that changes in the electrocardiogram in patients with biliary tract disease are variable and may be coincidental, and that it is not justifiable to speak of improvement of the cardiac condition as a result of biliary surgery on the basis of a single pre-operative and post-operative cardiogram, since serial tracing may show instability of the cardiographic pattern, especially of the T waves. *A. Schott*

The Changes in the Electrocardiogram Associated with Standing. D. Scherf and M. Schlachman. *Proc. Soc. exp. Biol., N.Y.*, **68**, 150–153, May, 1948.

Records in 80 male patients without evidence of organic heart disease were taken in the supine position, after standing for 1, 5, and 15 minutes, and again immediately upon resuming the supine position. To investigate the part played by the sympathetic nervous system 0·5 mg. of dihydrœrgotamine ("DHE 45") was given intravenously to 12 patients and records were again taken supine and erect when the drug effect was at its height. In 25 (31%) there were significant changes in the electrocardiogram on standing, but neither the kind nor the time of appearance of changes was uniform. In 4 temporary A-V rhythm was observed as a result of change of posture. In 11 out of 12 changes occurring immediately on standing could not be prevented by dihydroergotamine. It is concluded that the immediate and delayed changes in the cardiogram must be ascribed to different mechanisms. The former are due to the change of position of the heart and altered contact between the heart and neighbouring structures, the latter to the sympathetic nervous system acting on the heart directly or through the coronary arteries. *A. Schott*

A Clinical and Electrocardiographic Study of Paroxysmal Ventricular Tachycardia and its Management. G. R. Hermann and M. R. Hejtmancik. *Ann. intern. Med.*, **28**, 989–997, May, 1948.

In a heart that is damaged failure may be caused by sudden rapid rate. Most hearts in which ventricular tachycardia develops have been previously damaged by coronary disease or digitalis. A ventricular tachycardia is recognized by abnormally broad QRS complexes in the electrocardiogram along with an independent atrial rhythm.

Twenty patients with ventricular tachycardia are reported of whom 14 had coronary disease with or without infarction. The others had rheumatic heart disease, except for 2 in whom no organic disease could be found. Nine of the 20 were receiving digitalis at the onset of the attack. In 10 the heart rhythm reverted to normal on quinidine. Quinidine by mouth in a single oral dose produces a maximum concentration in the heart in about an hour, being eliminated in 8 hours. The largest dose used was a total of 5·2 g. in 24 hours. The method of Hepburn and Rykert of intravenous dosage is useful, 3·5 g. of quinidine sulphate in 500 ml. of 5% glucose intravenously at 100 ml. per hour. Once normal rhythm has been restored, quinidine should be continued by mouth for several days or weeks, the dosage being adjusted to prevent premature ventricular contractions. Morphine intravenously has also been used successfully, 10 to 40 mg., repeated after half an hour to 2 hours. Intravenous magnesium sulphate has also been used successfully. The prognosis is that of the underlying cardiac disease. In some cases achievement of a normal rhythm may not in itself prevent a fatal outcome.

J. McMichael

Experience with the Schemm Regimen in the Treatment of Congestive Heart Failure. A. A. Newman and H. J. Stewart. *Ann. intern. Med.*, **28**, 916–939, May, 1948.

The importance of a low salt intake in controlling œdema is now widely recognized. It has even been shown that, provided the salt intake is low, large amounts

of fluid may be taken without increasing cardiac œdema. Schemm carried this principle to the point of recommending a very high daily fluid intake with a diet low in sodium and yielding a neutral or acid ash residue. Thirty patients were admitted to hospital for trial of this regime. Failure to maintain a steady state of œdema during a control period led to the elimination of 21 patients from later analysis. In only 9 were the data satisfactory. The regime alone was without beneficial action. Five of the patients were quite unable to consume the large amounts of water recommended. Moreover, the diet was not liked by the majority of patients. After an adequate trial of the Schemm treatment, the condition in 7 patients was found to be much improved on a regime of restricted salt, limited fluid and frequent administration of mercurial diuretics. Analysing critically the charts in Schemm's papers, the authors do not think that diuresis was achieved on his regime. The only occasions on which the patients lost weight were when mercurials were given. *J. McMichael*

Aortic Stenosis. A Study of the Clinical and Pathologic Aspects of 107 Proved Cases. C. W. KUMPE and W. B. BEAN. *Medicine, Baltimore*, **27**, 139–185, May, 1948.

To facilitate the diagnosis of aortic stenosis the authors studied the clinical and pathological records of 107 post-mortem examinations, in which aortic stenosis with calcium deposits had been demonstrated in the absence of any other valve lesion.

The ages of the patients varied from 10 to 80 years, maximum incidence seventh and eighth decades, ratio males to females 3 to 1. A history of acute rheumatic fever was obtained in two-thirds. Thirty-four patients had chronic congestive failure, 10 had intermittent bouts of failure, and 19 gave a history of an abrupt onset of failure shortly before admission. Cardiac pain was present in only 9 and syncope in 4. The pulse rate was accelerated. Blood pressure was not characteristic. On auscultation the aortic second sound was usually absent or diminished, a systolic murmur was heard at the base in 83% with transmission to the neck in less than half. Basal diastolic murmurs were heard in one-third of the cases. Similar systolic and diastolic murmurs were heard at the apex. Basal thrills were felt in 33 cases, and though usually related in intensity to the degree of stenosis they were absent in several severe cases. Congestive failure was unusually refractive to treatment and was associated with much sweating. Cardiac pain differed from typical angina pectoris in its lack of radiation or radiation to the right, its advent after, rather than during, exercise and its resistance to nitroglycerin. It was more closely associated with severe aortic stenosis than with coronary arteriosclerosis. Death occurred suddenly in 21% of patients, usually after 5 to 30 minutes, in contradistinction to the instant death in some cases of myocardial infarction. Coronary arteriosclerosis was common and was associated with myocardial infarction in an appreciable number of cases. Arteriosclerosis was common in the abdominal and descending aorta but not in the ascending.

The condition was diagnosed clinically in only 24% of cases, signifying too great an acceptance of the classical triad of basal systolic murmur, thrill, and small, slow-rising pulse. *W. T. Cooke*

The Diagnosis of Mitral Insufficiency in Rheumatic Children. A. G. KUTTNER and M. MARKOWITZ. *Amer. Heart J.*, **35**, 718–726, May, 1948.

In order to assess the importance in rheumatic children of a loud blowing systolic murmur in the absence of demonstrable cardiac enlargement a comparison was made of the after-history of 144 children having such a murmur with that of 171 similar patients with potential and possible heart disease but with not more than a soft systolic murmur. The average follow-up period was 8 years (5 to 19 years). Those with the loud blowing murmurs were more susceptible to rheumatic fever as judged by the incidence of recurrences (63% had multiple attacks) than were the group with only potential rheumatic heart disease (31% had multiple attacks). Sixty-nine (48%) of those with "mitral insufficiency" developed organic heart disease, and 13 died of rheumatic infection and 7 of bacterial endocarditis. Only 22 (13%) of the patients with potential rheumatic heart disease developed it and none died.

These observations suggest that the diagnosis of mitral insufficiency, based on a loud blowing apical systolic murmur, is justified in children and carries a grave prognostic significance. *H. E. Holling*

Studies on the Coronary Circulation. III. Collateral Circulation of Beating Human and Dog Hearts with Coronary Occlusion. M. PRINZMETAL, H. C. BERGMAN, H. E. KRUGER, L. L. SCHWARTZ, B. SIMKIN, and S. S. SOBIN. *Amer. Heart J.*, **35**, 689–717, May, 1948.

Several methods of study were used in this investigation. Red blood cells labelled with radioactive phosphorus were injected into moribund patients shortly before death. At necropsy the heart was removed and the distribution of the radioactive cells was quantitatively determined by the Geiger counter and radio-autographs. Five hearts were studied, 2 normal and 3 from patients with myocardial infarction who died 4 days, 12 days, and 8 weeks after the onset of the attacks. Despite certain criticisms that may be advanced against these observations, the authors conclude that in arteriosclerotic hearts with myocardial infarction in living man there is a functioning collateral circulation which allows blood to enter all parts of a myocardial infarction, including the central portions.

Red cells labelled with radioactive phosphorus were injected into dogs at varying times after ligation of the anterior descending branch of the left coronary artery. At various intervals thereafter the hearts were stopped suddenly by freezing. The distribution of the radioactive cells was quantitatively determined. These observations lead the authors to conclude that: (*a*) Blood from collateral channels supplies the entire mass of ischæmic myocardium distal to a ligated coronary artery. (*b*) The ischæmic right ventricular myocardium supplied by the ligated artery is better nourished by collateral blood than is a similar portion of the left

ventricle. (*c*) The sub-epicardial portion of the ischæmic myocardium is better nourished than the sub-endocardial region. (*d*) The anastomotic blood continues to enter the ischæmic myocardium for at least 30 minutes after coronary artery occlusion. (*e*) The collateral blood supply to the ischæmic myocardium is an actively circulating one which supplies the entire ischæmic region.

The observations are thought to explain in man: (*a*) the rarity of infarction of the right ventricle; (*b*) the greater infarction of the sub-endocardial than of the sub-epicardial muscle; (*c*) the fact that infarcts are generally smaller than the mass of muscle supplied by the occluded vessel. *R. T. Grant*

The Effect of Occlusive Arterial Diseases of the Extremities on the Blood Supply of Nerves. Experimental and Clinical Studies on the Role of the Vasa Nervorum. J. T. ROBERTS. *Amer. Heart J.*, **35**, 369–392, March, 1948.

In a series of dogs the blood supply to nerves was interfered with in various ways—by ligation of the nutrient arteries, by stripping off the perineurium, by compressing and stretching the nerve, and by injecting air or particulate matter into the arteries. The degree of ischæmia was assessed by the results of inter-arterial injections of dye. The interference with blood supply was found to alter the function and structure of the nerves. Clinical studies in man showed that the blood supply to peripheral nerves may be reduced by similar processes in a number of conditions. Sensory and motor changes are closely related with the ischæmia of the nerves. A reflex arc for explaining referred pain on the basis of neural ischæmia is proposed. *R. T. Grant*

The Effect of Arteriosclerosis on the Dynamics of Hypertension in the Aged. A Preliminary Clinical and Pathological Study of 150 Cases. F. D. ZEMAN and B. M. SCHWARTZ. *J. Gerontol.*, **3**, 40–47, Jan., 1948.

The authors report clinical and necropsy findings in 150 unselected elderly subjects, observed for periods of from a few days to 20 years. The maximum number of blood-pressure estimations on any patient was 57, the minimum 1. Blood pressure, taken during routine medical visits, was classified as: normal (150/90 mm. Hg.); systolic hypertension (over 150/90 or under); systolic and diastolic hypertension (over 150/over 90). On this basis patients were placed in 4 main groups: (1) (*a*) normal, 12%; (*b*) mainly normal, occasional variations, 11%; (2) (*a*) every systolic reading raised, with every diastolic reading normal, 13%; (*b*) most systolic readings raised, with most diastolics normal, 26%; (3) (*a*) both pressures raised (except terminally), 17%; (*b*) occasional variations of this, 11%; (4) the remaining 9% in whom readings were variable. One-third of all males had evidence of coronary occlusion. In females the incidence was much lower. Arteriosclerosis of the aorta was present in all groups, but most severe in (2). Renal arteriosclerosis was most marked in group (3). Cerebral accidents became commoner as blood pressure rose.

Necropsy showed that peripheral resistance increases with age, and this factor, with decreased aortic elasticity, gives rise to systolic hypertension, which may, however, really be a modified systolic-diastolic hypertension. Pure systolic hypertension occurs when decrease in aortic elasticity is greater than increase in peripheral resistance. The authors stress the need for consideration of cardiac, cerebral, and renal function before a blood pressure level is taken as indicative of cardiovascular disease; a "normal" blood pressure in old age is produced by interaction of the above variables, and does not afford evidence of normality. *Morag L. Insley*

Heart Disease in Pregnancy. D. J. MACRAE. *J. Obstet. Gynaec. Brit. Emp.*, **55**, 184–198, April, 1948.

Heart disease is a common and serious complication of pregnancy. There was an incidence of 0·8% in 29,713 patients attending Queen Charlotte's Maternity Hospital from 1937 to 1946 inclusive, with a mortality of 3%. In the series reviewed 11% of the maternal deaths were due to heart disease.

Pregnancy increases the work of the heart to such an extent that a damaged heart may be unable to bear the extra strain. The most valuable single method of assessing cardiac function in these cases is by determining the response to the routine of daily life. The classification adopted by the New York Heart Association is recommended: Group 1: no limitation of normal active life. Group 2: slight limitation, producing breathlessness at the end of effort. Group 3: definite limitation necessitating resting two or three times while climbing stairs. Group 4: heart failure at rest. Auricular fibrillation is a most serious complication; in the series reviewed, of 7 patients with fibrillation 3 died. Once decompensation occurs in pregnancy the chance of its recurring in subsequent pregnancy is great, and termination requires consideration. This is also advised for Group 4 patients and those in Group 3 who in the early months fail to respond to medical treatment. This consists of maximum rest, with at least 2 hours in the afternoon and 12 at night. Weekly examinations are advised throughout pregnancy. Intercurrent infection should be treated seriously, and admission to hospital for a week of observation at the twenty-eighth week is advised, and again for the week preceding delivery. Vaginal delivery is the method of choice with adequate sedatives and forceps in the second stage. In Group 4 no obstetrical treatment should be attempted until the patient is thoroughly rested and digitalized.

Cæsarean section was performed on 21 patients in this group, but its role in patients with heart disease is strictly limited, as for example in heart disease with associated disproportion. The need for sterilization should not be used as an argument in its favour.

In these 225 patients with heart disease, 124 were suffering from mitral stenosis, 23 had an associated aortic incompetence and 13 had congenital lesions, of which the most serious was coarctation. In such cases pregnancy should be avoided, but if it does occur Cæsarean section is advised as the method of delivery.

The avoidance of infection in the puerperium and the

importance of adequate rest are stressed; breast-feeding was not permitted in Group 3 and 4 patients. As most of the fatalities occurred in the puerperium the difficulty of giving a prognosis in the early stage of pregnancy is mentioned. Patients may easily pass from one group to the other during the course of pregnancy or labour, but the prognosis has greatly improved with better antenatal and intra-partum care. *J. Stallworthy*

The Electrocardiogram in Mitral Stenosis with Special Regard to its Development. A Study of 100 Cases. H. RASMUSSEN and G. NYHUS. *Acta med. scand.*, **129**, 446–471, Jan. 27, 1948.

The authors state that their object in this study was to seek electrocardiographic peculiarities, other than those usually described, that might be of value in diagnosis. One of the authors believed that he had observed, in a previous study of the cardiogram in diseases of the left side of the heart, progressive changes leading to a left bundle-branch block. It was therefore possible that similar changes might be found in disease affecting chiefly the right side of the heart.

Serial studies of the electrocardiograms of 100 patients and of the size of the heart show that the cardiographic changes and increase in the size of the heart appear simultaneously during the progress of the disease. Retardation of the impulse to the right ventricle is held to be decisive for the cardiographic development. The retardation may cease in the stage with low R_1 or may progress until a maximal right ventricular retardation curve appears, ending with right bundle-branch block. The latter occurs in the same proportion of cases as does left bundle-branch block in left heart disease.

Donald Hall

The Arterial Oxygen Saturation in Cyanotic Types of Congenital Heart Disease. G. E. MONTGOMERY, J. E. GERACI, R. L. PARKER, and E. H. WOOD. *Proc. Mayo Clin.*, **23**, 169–176, April 14, 1948.

This communication deals with the application of the oximeter to measurements of oxygen saturation of the blood. The oximeter used was that described by Millikan in 1942 and fitted with a photo-electric colorimeter. The oxygen saturation of the blood is measured directly by fitting the apparatus on to the human ear. The first study was concerned with the oxygen saturation of the blood in 25 patients with congenital cardiac defects of the cyanotic group; 19 healthy subjects served as controls. Readings of oxygen saturation of arterial blood were taken and the effects of breathing 100% oxygen, change of posture, and exercise were particularly noted.

With the patients breathing pure oxygen the arterial saturation increased by from 2 to 16 points %, while in the normal controls this was only increased by from 1 to 5 points. The effect of walking less than 2 miles an hour diminished the arterial oxygen saturation by from 3·5 to 19 points % in the patients while the maximum in the normals was only 2 points %.

Interesting data are given on 1 patient with the tetralogy of Fallot, who, when exercised on the treadmill for 5 minutes at 1·7 miles per hour, showed a fall of arterial oxygen saturation from 80% to about 50%; in a normal subject in the same circumstances the oxygen saturation increased slightly.

The second part of the paper deals with the determination of arterial oxygen saturation at rest and during exercise in 8 cyanotic patients. This was done both by the Van Slyke method and by oximeter readings. The results were then compared and it was found that the average decrease in saturation on exercise was 19·0% by the Van Slyke method, while the simultaneous oximeter readings indicated a 13·3% decrease. Thus the discrepancies are large, but, as the authors say, "the instrument is still of considerable value in estimating the degree of disability of a patient and in judging the efficacy of corrective surgical procedures in such patients." Suitable data are given in support of these statements.

A. I. Suchett-Kaye

Electrokymographic Studies of Asynchronism of Ejection from the Ventricles. Normal Subjects and Patients with Bundle Branch Block. G. F. ELLINGER, F. G. GILLICK, B. R. BOONE, and W. E. CHAMBERLAIN. *Amer. Heart J.*, **35**, 971–979, June, 1948.

Analyses of electrokymograms indicate that, in normal subjects, asynchronous ventricular ejection is more frequent than synchronous ejection. In patients with left bundle-branch block ejection from the right heart and in patients with right bundle-branch block ejection from the left heart precedes that of the opposite side by a significantly longer time than in normal subjects.

R. T. Grant

The Effect of Spinal Anesthesia on the Renal Ischemia in Congestive Heart Failure. R. MOKOTOFF and G. ROSS. *J. clin. Invest.*, **27**, 335–339, May, 1948. 12 refs.

High spinal anæsthesia caused no significant changes in renal plasma flow and glomerular filtration rates in 11 cases of chronic congestive heart failure. The ischæmia with vasoconstriction associated with this condition cannot be attributed, therefore, to neurogenic stimulation following a fall in cardiac output.

E. F. McCarthy

Oxygenation Studies in Congenital Pulmonary Stenosis. An Application of Recording Oximetry in the Evaluation of Cardiorespiratory Function. G. GULLICKSON, J. O. ELAM, H. HAMMOND, J. R. PAINE, and R. L. VARCO. *Amer. Heart J.*, **35**, 940–947, June, 1948.

In 10 cases of the tetralogy of Fallot the arterial oxygen was estimated by a recording oximeter before, during, and after operation for the establishment of a systemic-pulmonary arterial shunt. The chief findings were that: (1) a rise in arterial oxygen saturation occurs practically immediately on the establishment of the shunt; (2) as a result of the operation the saturation time upon administration of 100% oxygen is greatly shortened.

R. T. Grant